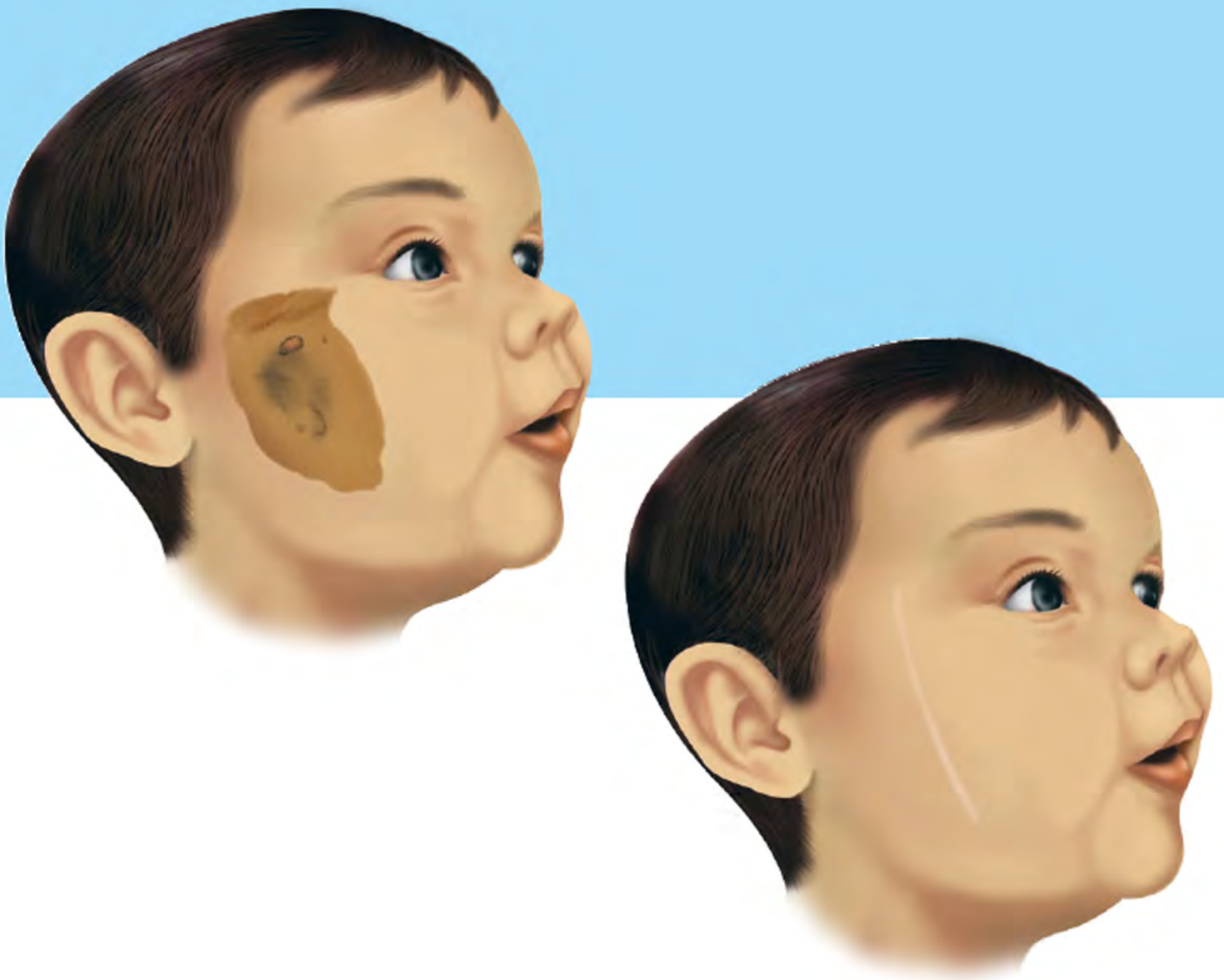


Pediatric Plastic and Reconstructive Surgery

Arin K. Greene

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Pediatric Plastic and Reconstructive Surgery

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Preface

The field of pediatric plastic surgery is evolving. Plastic surgeons who sought specialty training in pediatric conditions classically fulfilled a “craniofacial” fellowship often focused on craniofacial syndromes, craniosynostosis, and cleft lip/palate. This paradigm has been changing and many plastic surgeons interested in treating children seek broader education. Plastic surgeons working in a children’s hospital understand that “craniofacial” problems comprise only a portion of pediatric plastic surgical diseases.

The goal of this book is to provide a comprehensive reference for pediatric plastic surgeons, fellows, residents, medical students, and allied health professionals. The text was designed to be clinically oriented, rather than focused

on history or research. The book was written to be a single volume, easy-to-read resource that highlights principles.

Authors and section editors were chosen because of their expertise on their respective topic(s). The book was organized by what I consider to be the major categories of pediatric plastic surgical diseases: Craniofacial Disorders, Cleft Lip and Palate, Soft Tissue Deformities of the Head and Neck, Trunk and Lower Extremity, Upper Extremity, and Integument. In addition to serving as a roadmap for the treatment of patients, my hope is that the text can be used as a resource for more in-depth study of a subject, teaching, and/or research. Ideally, it will stimulate readers to innovate and improve patient management.

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1 Craniosynostosis

Gary F. Rogers and Stephen M. Warren

Summary

Craniosynostosis is the premature fusion of one or more cranial sutures. It can occur in isolation (non-syndromic) or in association with a syndrome (syndromic), and can involve a single or multiple cranial sutures. Patients with non-syndromic single suture fusions have normal intelligence, but may display subtle learning disabilities. Patients with a syndromic diagnosis and/or multiple suture fusions tend to have variable degrees of cognitive impairment. The sagittal suture is the most commonly (40 to 55%) fused suture followed by the coronal (20 to 25%), metopic (5 to 15%), and lambdoid (1 to 5%) sutures. This condition has two primary consequences: a characteristic change in cranial shape and, more importantly, potential restriction of cerebral growth and development. The morphologic effects of suture fusion are related to the location and number of fused sutures, and the age at which the fusion(s) occurred. Diagnosis is typically made by physical examination (i.e., the shape of the head) and/or computed tomography. The general goals of treatment are to normalize cranial shape and intracranial volume, but other considerations (e.g., respiration, ophthalmologic, facial aesthetics, musculoskeletal) may require additional intervention. Treatment options to address the craniosynostosis include: minimally-invasive procedures (endoscopically-assisted strip craniectomy (SC), SC+spring mediated distraction), open craniotomy procedures (Pi, open release), cranial distraction, and open cranial remodeling with osseous stabilization. Results of treatment vary according to the presence/absence of an associated syndrome, age at procedure, type of procedure used, duration of follow-up.

Keywords: Craniosynostosis, intracranial hypertension, syndrome, cranial suture

1.1 Introduction

Craniosynostosis is defined as the premature fusion of one or more cranial sutures. This condition occurs in an estimated 1 in 2,000 to 2,500 live births, and can occur as a solitary finding (termed nonsyndromic craniosynostosis) or in conjunction with pathologically associated physical anomalies (syndromic). The majority (80–95%) of affected patients have a single fused suture and no concurrent syndrome. Of the patients with multiple suture fusions, approximately 75% have an associated syndrome. The sagittal suture is the most commonly involved (40–55%), followed by the coronal (20–25%), metopic (5–15%), and lambdoid (1–5%) sutures. Some recent reports document a rise in the incidence of isolated metopic fusion and place it second behind sagittal synostosis; this finding is not universally reported and may reflect diagnostic variability between providers and centers and not a true rise in pathoanatomy. Fusions of other sutures (e.g., squamosal and frontosphenoidal) are much less common.

Craniosynostosis has two primary consequences: a characteristic change in cranial shape and, more importantly, potential restriction of cerebral growth and development. The morphologic effects of suture fusion are related to the location and

number of fused sutures and the age at which the fusion(s) occurred. Most documented forms of craniosynostosis occur *prenatally*, and malformation of the craniofacial skeleton is evident at birth. However, there are rare examples of abnormal sutural fusion that occur after birth; in such instances, the changes in cranial shape may be subtle and difficult to diagnose.

1.2 Cranial Development, Anatomy, and Pathoanatomy

Cranial sutures and fontanelles are the mesenchymes that persist between the calvarial plates. The major cranial sutures are the metopic, sagittal, coronal, and lambdoid sutures (► Fig. 1.1). Minor cranial sutures include the temporosquamosal, frontonasal, sphenoethmoidal, and the frontosphenoidal sutures. The cranial sutures have two primary functions: to permit cranial deformation or molding during parturition so that the infant can pass through the pelvis and to allow the cranium to accommodate accelerated brain development early in life. As the brain grows, tension across the suture increases and stimulates bone deposition along the edges of the calvarial plates. Thus, cranial growth occurs in response to brain growth. Once brain growth is completed, the cranial sutures serve little purpose and eventually close. Conventional wisdom suggests that the cranial sutures close in childhood, but histologic sections suggest that true bony fusion occurs much later.

The complex biological activity of the cranial sutures is still being elucidated. Sommering (1800) was the first to describe the anatomy of the cranial sutures and to propose that premature fusion of the suture could alter cranial shape. Rudolph Virchow (1851) expanded his work and posited that cranial suture fusion (or patency) was independent of the perisutural environment. He further hypothesized that premature suture fusion resulted in compensatory skull growth parallel to the fused suture and a decrease in growth perpendicular to the suture (Virchow's law; ► Fig. 1.2). Although Virchow's law is the foundation for clinical diagnosis of craniosynostosis, the understanding of sutural biology has changed dramatically. Beginning with the work of various researchers, including Park and Powers (1920), Van der Klaauw (1946), and Moss (1959), attention began to focus on the influence of the dura mater on suture activity, patency, and closure. Numerous *in vitro* and *in vivo* models strongly suggest that the underlying dura mater mediates suture activity through temporal and spatial production of growth factors (e.g., fibroblast growth factor [FGF]-2) and cellular elements (e.g., osteoblastic cells) to the overlying osteogenic fronts and suture mesenchyme.

Genetic methods have now identified more than 100 mutations that are involved in the pathogenesis of craniosynostosis, including *TWIST*, *NELL-1*, *MSX-2*, *GLI3*, and fibroblast growth factor receptor (*FGFR*)-1 to -3. These are covered more extensively in Chapter 2 on craniofacial syndromes. The mechanism through which these mutations cause craniosynostosis is still being elucidated. Craniosynostosis appears to affect facial

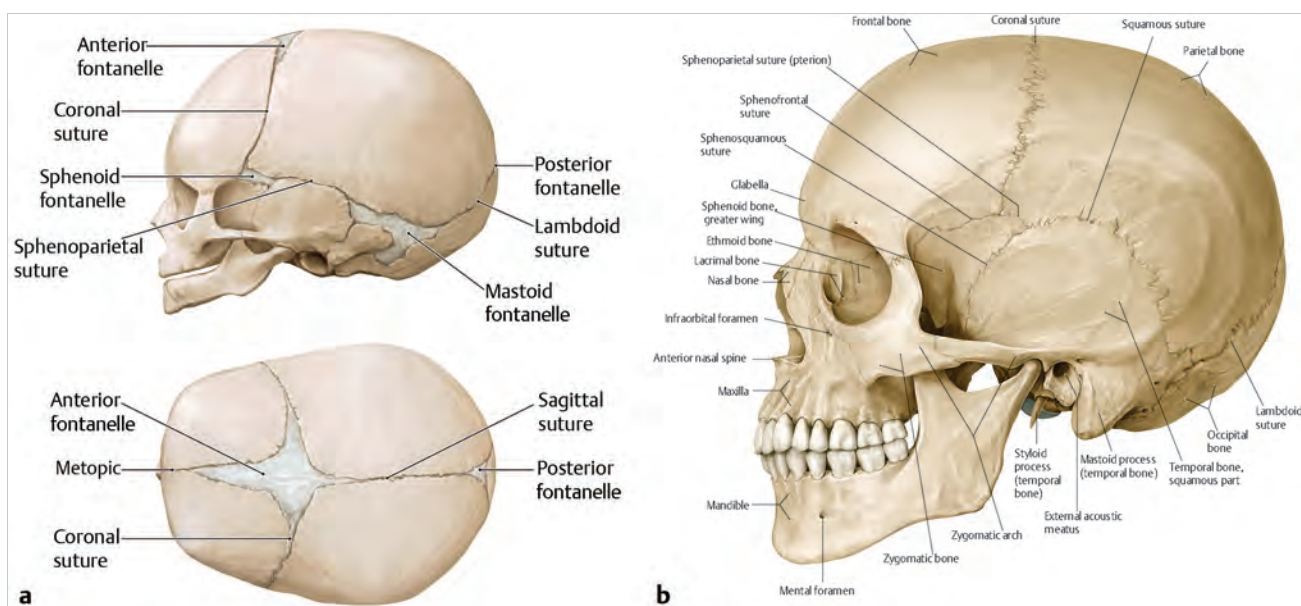


Fig. 1.1 Cranium. (a) Vertex view. (b) Lateral view. (From THIEME Atlas of Anatomy, Head and Neuroanatomy, © Thieme 2010, illustration by Karl Wesker.)

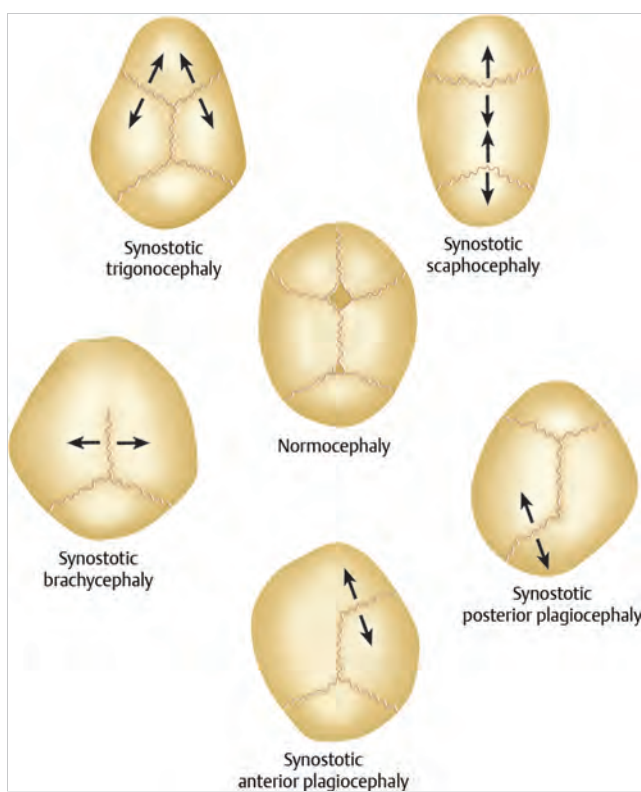


Fig. 1.2 Virchow's law.

growth through a secondary cascade of growth impairment that extends from the cranial base into the facial skeleton. Findings by Mooney and others suggest that the calvarial dysmorphology can drive the basicranial and midface changes. The controlling influence of genetics cannot be minimized, but epigenetic influences and mechanotransduction can modulate

genetic expression and patient phenotype. There are well-documented examples of individuals who possess genes associated with certain syndromic forms of craniosynostosis (e.g., *FGFR-3 Pro250Arg* and *TWIST*) and yet lack any evidence of cranial fusion or the phenotypic features of the underlying syndrome.

1.3 Classification

Craniosynostosis can be generally classified based on both the number of involved sutures (single vs. multiple) and the presence or absence of an associated syndrome (nonsyndromic vs. syndromic). In general, most patients (~85%) with single-suture craniosynostosis have no identifiable associated syndrome (nonsyndromic, single-suture craniosynostosis), whereas most patients (~75%) who carry a syndromic diagnosis have fusion of two or more sutures (syndromic, multisuture craniosynostosis). However, it is incorrect to use the terms single-suture and nonsyndromic or multisuture and syndromic interchangeably. There are many examples of patients with isolated sutural fusions who have an associated syndromic diagnosis (syndromic, single-suture craniosynostosis) and patients with multisutural fusion that have no identifiable syndrome (nonsyndromic, multisuture craniosynostosis). Approximately 25% of patients with unilateral coronal synostosis have an associated molecular/syndromic association: Muenke's syndrome (*FGFR-3 Pro250Arg*; most common), Apert's syndrome (*FGFR-2*), Saethre-Chotzen syndrome (*TWIST*) and craniofrontonasal syndrome (*EFNB1*). Similarly, an estimated 28% of patients with metopic craniosynostosis have an associated syndrome or chromosomal deletion/transposition.

Although no classification system is perfect, this simple method captures two of the most important prognostic variables of craniosynostosis—the risk of intracranial pressure (ICP) and whether or not there is an associated syndromic diagnosis. Elevations in ICP are directly correlated with the number of

fused sutures. Marchac and Renier measured the ICP in 121 patients with craniosynostosis with the help of an epidural sensor. They detected elevated ICP in 42% of patients with multiple-suture involvement and in 7 to 13% of patients with single-suture involvement. They noted a decrease in ICP in patients who underwent cranial release. Similar results have been observed by other authors.

The presence or absence of a syndromic diagnosis greatly affects a patient's overall prognosis in terms of cognitive and functional capabilities. Most of the genes linked to the various forms of syndromic craniosynostosis (e.g., *FGFR-1 to -3*) are expressed in multiple tissues during embryogenesis, including neural tissue. The effect of a mutated gene during critical periods of development may, and very likely does, have a more profound effect on neurocognitive development than growth restriction produced by sutural fusion later in childhood. Given the importance of the genetic influences, it is paramount that every child with craniosynostosis undergoes genetic evaluation and testing for commonly associated genetic mutations.

1.4 Diagnosis

Physical examination remains the gold standard for diagnosing craniosynostosis. Most forms of craniosynostosis involve fusion of only a single suture and manifest as a characteristic cranial shape at birth. There are rare forms of craniosynostosis, such as those involving the minor cranial sutures (e.g., frontosphenoidal and squamosal), multiple contiguous or noncontiguous sutures, and latent (postnatal) fusions, that do not fall neatly into the standard phenotypic patterns promulgated by Virchow and are challenging to diagnose by phenotype alone. Most craniofacial surgeons choose to obtain a confirmatory computed tomographic (CT) scan in all patients, but others rely exclusively on the clinical findings for isolated forms of single-suture craniosynostosis. The primary advantage of obtaining a CT before embarking on surgical intervention is to identify any intracranial anomalies (hydrocephalus, arachnoid cysts, etc.) that may also require treatment. In addition, some centers now use virtual surgical planning algorithms that necessitate CT imaging. The disadvantages include the possible need for a general anesthetic and exposure to ionizing radiation. There is evidence that suggests that early exposure to either of these can have possible adverse effects, but the risks are extremely low and must be balanced against the need for clinical information potentially derived from the CT. It is the authors' current practice to obtain CT imaging on all patients, regardless of the fusion type.

1.5 Functional Aspects

1.5.1 Elevated Intracranial Pressure

The possibility of elevated ICP ranks as the most definitive and objective reason to consider surgical intervention in a patient with craniosynostosis. Prolonged elevated ICP can impair neurocognitive function, lead to blindness, and exacerbate or induce Arnold-Chiari malformation. As noted earlier, the risk of this problem rises geometrically with the number of fused sutures. The most common reason for this finding is a mismatch in shape and volume between the cranium and the brain,

but this can be exacerbated by other conditions. Sleep apnea resulting from midfacial retrusion can induce episodic nocturnal elevations in ICP secondary to the dilating effects of hypercapnia on the cerebral vasculature. Another potential cause is venous hypertension resulting from stenosis or complete closure of the sigmoid/jugular sinus complex.

It is important to recognize that although most patients with craniosynostosis do not have elevated ICP, the absence of reliable noninvasive methods of determining ICP (i.e., fundoscopic examinations, radiographic changes, and symptoms) and impracticality/morbidity of repeat invasive monitoring make observation a risky proposition. The gold standard for detecting elevated ICP is direct monitoring, which involves placing a transcranial monitor into the cerebral parenchyma or ventricles and monitoring pressure for 12 to 24 hours. This procedure requires an anesthetic and has some risk of intracranial bleeding or cerebrospinal fluid leak. Other methods of assessing ICP such as epidural monitoring and lumbar puncture have fewer complications, but the accuracy of these techniques is questionable. Intracranial pressure readings tend to fluctuate significantly with patient position, activity, blood pressure, and sleep, and the most meaningful results are obtained when patients are monitored for a period of time, usually overnight. Significant elevations (> 20 mm Hg) have been considered an absolute indication for intracranial expansion. However, interpretation of the significance of borderline pressure elevations (15–20 mm Hg) has been more problematic, and there is little consensus, even among neurosurgeons.

Direct ICP monitoring is invasive and rarely used for routine screening. Moreover, the measurement is only a snapshot in time: a normal pressure measurement early in life does not imply that it will remain so as the brain continues to grow. Consequently, many surgeons resort to noninvasive, but less reliable, indicators of ICP. Conventional clinical symptoms of acute ICP elevation, such as headache, somnolence, and dizziness, are often lacking, even in severely affected children. Papilledema and subsequent optic atrophy are strongly suggestive of elevated ICP but have limited sensitivity in children younger than 8 years. As optic atrophy progresses, the disk becomes pale, the capillaries and hyperemia disappear, and significant secondary arteriolar narrowing occurs (► Fig. 1.3). Reducing ICP can reverse early changes, but more advanced degeneration may be permanent. Radiographic evidence suggestive of elevated ICP includes loss of subdural space, often with effacement of the basal cisterns and vertex sulci, ventricular compression, and scalloping of the cranial endocortex. This latter finding has been termed the “copper beaten” skull and can be visualized on both conventional radiography and CT (► Fig. 1.4). It is a late finding caused by pressure remodeling of the inner table of the skull by the cerebrum. The correlation of this finding with elevation in the ICP has been questioned.

1.5.2 Hydrocephalus

Hydrocephalus is an infrequent finding in patients with craniosynostosis. When it occurs, it is more common in patients with Crouzon's syndrome but can also occur in other syndromes. It is extremely rare in patients with nonsyndromic, single-suture fusion. Findings of hydrocephalus may include ICP elevation and the presence of enlarged or enlarging ventricles. CT scans

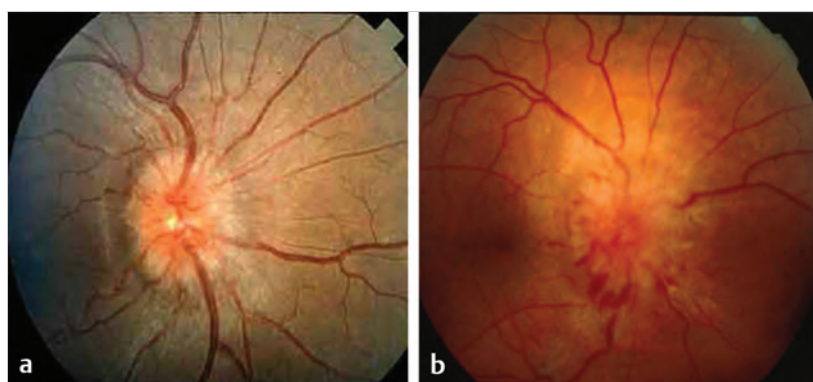


Fig. 1.3 Optic disc. (a) Normal. (b) Papilledema with blurring of the disc edges.

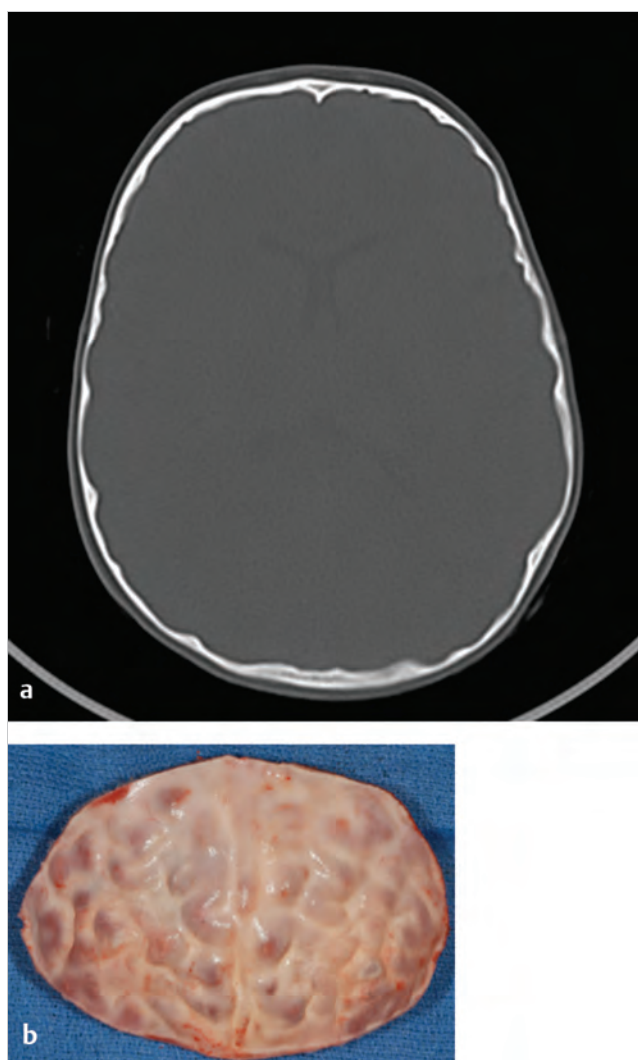


Fig. 1.4 Endocortical erosions. (a) CT findings. (b) Intraoperative view of the frontal bone with endocortical scalloping.

provide a reasonable noninvasive method of assessing ventricular size. Interestingly, the authors have observed cranial suture fusion following rapid decompression of hydrocephalus. This postnatal iatrogenic form of craniosynostosis is rare but has been reported.

1.5.3 Mental Impairment

Children with craniosynostosis can exhibit neurocognitive delay and/or learning disabilities. Risk factors include the presence of an associated syndrome or genetic mutation, concurrent ICP elevation, hydrocephalus, prematurity, and family history. Some authorities point to the deleterious effects of early cerebral growth restriction as the primary cause. However, several large investigations have failed to demonstrate that cranial remodeling or volume expansion significantly alters neurocognitive parameters. It is also likely, especially in those with a syndromic diagnosis, that the underlying genetic process that initiated premature sutural fusion also adversely impacted the development of the central nervous system. Patients with single-suture fusion and no associated syndrome generally have near normal intelligence but may exhibit subtle learning disabilities or delayed speech acquisition. In contrast, patients with an associated syndrome have a significantly higher incidence of cognitive delay than in the general population. The degree of cognitive impairment is loosely correlated with the type of syndromic diagnosis, but there is wide variability.

1.5.4 Visual Abnormalities

Ocular anomalies are surprisingly common in patients with craniosynostosis. Hypertelorism, exorbitism, strabismus, and proptosis are observed in many syndromic forms of bilateral coronal craniosynostosis, especially in those with significant midface hypoplasia. These findings are secondary to decreased orbital depth and widening of the ethmoidal air cells and can lead to corneal exposure and ulceration. Hypotelorism and strabismus can be associated with metopic synostosis. Patients with unilateral coronal synostosis have elevation of the lesser and greater sphenoid wings on the side of the fuse suture (harlequin deformity) that results in strabismus and ocular torticollis (head tilt to unfused side) in nearly 80% of affected patients. In addition, the contralateral orbital roof is depressed and 55% of patients have astigmatism. Patients with Saethre–Chotzen syndrome (*TWIST* mutation) demonstrate upper eyelid ptosis. Many of these manifestations are disfiguring and some can threaten vision. Patients with strabismus or nonconjugate gaze can develop decreased vision (disuse amblyopia) if the visual axis disturbance is not corrected. Strabismus and amblyopia can occur in up to 40% of patients with syndromic craniosynostosis, but these findings are less common in nonsyndromic

patients. Patching of one eye and operative balancing of the extraocular muscles are the mainstays of treatment.

1.6 Treatment

1.6.1 Team Approach

Management of craniosynostosis can be complex and is best accomplished with coordinated, interdisciplinary care. This is the standard approach to care in most craniofacial centers, and the team comprises professionals from the following disciplines: anesthesiology, craniofacial surgery, genetics, hand surgery, intensive care, neurosurgery, nursing, ophthalmology, orthodontics, pediatrics, pediatric dentistry, prosthodontics, psychology, radiology, social work, and speech/language pathology. Given the number of specialists involved in a comprehensive craniofacial program, organizing and maintaining an experienced and well-coordinated team is challenging, especially given the financial pressures of “health care reform.” Moreover, reimbursement issues are progressively limiting access to certain specialists, diagnostic tests, and treatment options. However, such a comprehensive approach to care is one of the most critical elements in achieving the best overall outcome for the patient.

1.6.2 Timing of Surgical Intervention

The optimal timing for surgical treatment of craniosynostosis is debatable. One approach is to operate as soon as possible, in order to halt further progression of secondary craniofacial changes, allow normal brain expansion, and capitalize on the ameliorating effects of brain growth on overall skull shape. There is a higher likelihood that a young infant will spontaneously ossify any bone defects created during the operation. Moreover, some recent investigations suggest that early correction yields a better cognitive outcome compared with later surgery. However, there is a higher risk of anesthetic-related complications in infants younger than 1 year, and some authors have observed a greater need for revision in patients undergoing open remodeling procedures before 6 months of age. Anesthetic-induced neural apoptosis, or cellular death, is also a concern in very young infants who undergo large intracranial procedures. Such factors support deferring larger procedures until the infant is older, usually 9 to 12 months of age. Each of these considerations should be weighed in light of the *type* of procedure to be performed and the risks of delaying treatment.

1.6.3 Type of Procedure

There is considerable variation in treatment of craniosynostosis from one center to another. Operative alternatives in order of increasing invasiveness include endoscopic-assisted extended strip craniectomy with postoperative cranial orthosis (endoscopically assisted suturectomy with helmet therapy [EAS+HT]), suturectomy with spring mediated distraction, cranial osteotomies with internal screw distraction devices, and open partial or total cranial vault remodeling. The choice of treatment depends on many factors, including, but not limited to, the patient's age, the location and number of sutures fused, the

presence of an associated syndrome, severity of pathology, and surgeon/center experience.

Minimally invasive techniques offer less morbidity than larger open procedures but are generally used only in younger patients. These methods, which include EAS+HT and spring-mediated distraction, require very small skin incision(s), and the limited dissection can be facilitated by using an endoscope. The fused suture is removed along with strip of the adjacent bone, the width of which can vary from one to several centimeters (► Fig. 1.5). Although some authors advocate removing a wide segment of bone, there is no clear evidence that this strategy improves outcomes. Moreover, the risk of a persistent bone defect is higher than with more limited resections. The addition of lateral barrel staves has also been touted as advantageous, but there is again no evidence to suggest the superiority of this approach. Suturectomy is typically accompanied by postoperative helmet orthotic to guide the cranial expansion or intraoperative springs placed between the bone edges to facilitate expansion of the craniectomy gap. Although some authors question the necessity of these adjuncts, it is the authors' opinion that suturectomy without some adjuvant treatment method yields a suboptimal outcome. EAS+HT relies solely on rapid brain expansion to improve cranial shape, and, consequently, this technique is best limited to very young infants (usually <4 month of age). The orthotic is worn for 4 to 6 months, and usually two or more helmets are required to complete the treatment. In contrast, spring-mediated distraction does not depend on brain growth and has a wider window of effectiveness, up to 8 months, and can allow more expansion of the bone (► Fig. 1.6). A second minor procedure is required to remove the spring. Both approaches have minor blood loss (50–100 mL), short hospitalization (0–1 days), and very low

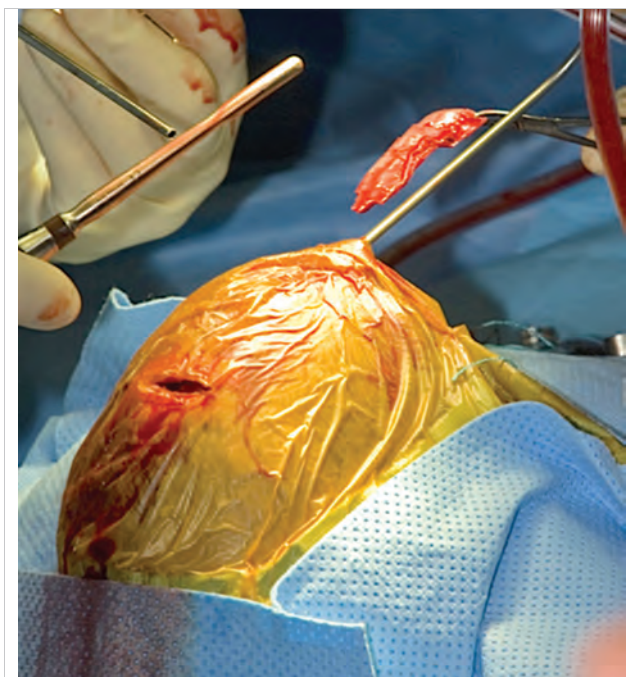


Fig. 1.5 Endoscopically assisted suturectomy with helmet therapy; minimally invasive removal of the sagittal suture.

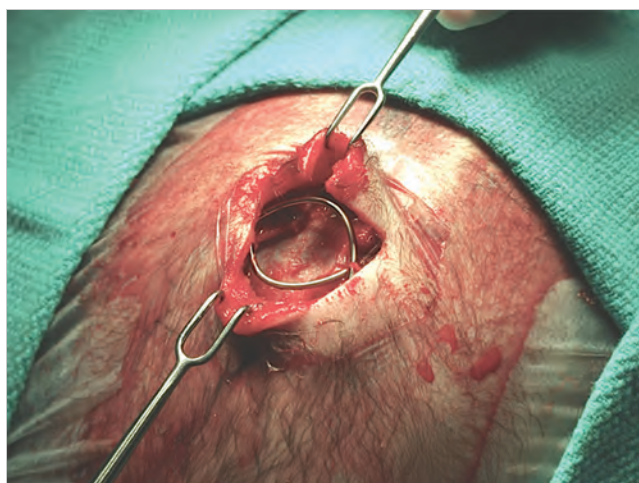


Fig. 1.6 Spring-mediated distraction; spring insertion for sagittal synostosis. (Image courtesy of Lisa David, MD.)

transfusion rates. The safety and effectiveness of these techniques are excellent for most types of single-suture and bilateral coronal craniosynostosis, but they should be used with caution in patients with complex multisuture fusions.

Distraction of the cranium is a newer technique that has shown promise for certain types of craniosynostosis. Distraction has now been applied to virtually every form of craniosynostosis, though more commonly in the posterior cranium in patients with bilateral coronal synostosis (early decompression) and to perform monoblock advancement (concurrent midfacial and frontal advancement). It can be used theoretically in patients of all ages and allows large osseous movements that would be difficult, if not impossible, to achieve with the minimally invasive or even open cranial remodeling procedures. Because this is a gradual process, ossification of the bone gaps is quite good. The procedure does require an open dissection to perform the craniotomy and place the distractors and a second procedure to remove the device. Moreover, a recent meta-analysis, including 11 studies of posterior cranial vault distraction, found that this technique had complication rates between 12 and 100% (average 30%), much higher than most other techniques used to correct craniosynostosis; these complications included cerebrospinal fluid leak, infection, exposure of the implants, and implant failure.

Open cranial remodeling techniques remain the gold standard in most craniofacial centers throughout the world. Promulgated by Tessier, these procedures require an open approach, typically using a coronal incision, and removal of various cranial bone segments. The bone is then manually remodeled and/or rearranged, repositioned, and stabilized. Most surgeons use micro- or miniplates and screws to secure the advanced bone segments. Early plating systems were made of titanium, but these plates and screws were noted to “migrate” intracranially when used in infants. Although no harmful effects were reported, most surgeons use resorbable plate fixation in young children (<2 years of age) when it is feasible. All open procedures have more blood loss (200–400 mL), longer operative times (2–5 hours), and longer periods of hospitalization than their minimally invasive counterparts. As noted earlier, some surgeons defer operative intervention until the infant is closer

to 1 year of age to reduce potential risks. However, open cranial remodeling is very safe and effective if done properly and in centers with an experienced support team.

1.7 Perioperative Considerations

There are two primary goals in operative management of craniosynostosis: to obtain the best possible anatomic outcome (i.e., sufficient intracranial volume and improve/normalize cranial shape) and patient safety. Consequently, operative treatment of craniosynostosis should only be done by experienced craniofacial teams in higher-volume centers. Inexperienced surgical and/or anesthetic teams not only risk producing a suboptimal anatomic correction (and a higher rate of reoperation) but also elevate the likelihood of an adverse event. This is especially true in syndromic patients or those with comorbid conditions.

Excessive blood loss and inadequate resuscitation are the principal causes of serious morbidity and death in open cranial vault surgery and mandates careful attention. The anesthesia team must constantly assess the blood loss (calculated) and administer blood products and intravenous (IV) fluids well before the patient experiences significant physiologic manifestations. Attempt to “catch-up” from inadequate resuscitation is not advised and puts the patient in significant peril. Larger open procedures usually necessitate placement of an arterial line, at least two peripheral IV lines (in case one fails during the procedure), and a Foley catheter; some also place a central line, although we have not found this necessary in most patients. Most experienced centers employ strategies to reduce blood loss, including meticulous surgical technique, application of bone wax to bleeding bone edges, and pharmacologic modalities such as tranexamic acid (TXA); TXA has been shown to significantly reduce blood loss in craniofacial procedures. Various blood salvage techniques have also been advocated, although they only reduce allogeneic transfusion risk and do not mitigate the risk of coagulopathy and hemodynamic instability associated with the blood loss.

Precordial Doppler monitoring can help identify air embolism before the onset of a hemodynamic emergency. Air embolus is most likely to occur when the patient is inadequately resuscitated and when bone is being cut or removed. If there is suspicion of air embolus, the operative site should be covered with a wet towel (or the scalp redraped) and the head of the bed should be dropped below the level of the heart until the threat has dissipated. Lastly, the use of perioperative IV steroids has been shown to dramatically reduce swelling, pain, and hospitalization; dexamethasone at a dose of 3.0 mg/kg before incision and then every 8 hours tapered over 3 days works very effectively.

1.8 Clinical Findings and Management of Specific Types of Craniosynostosis

1.8.1 Sagittal Synostosis

The sagittal suture is the most likely to fuse prematurely and comprises 40 to 55% of all forms of craniosynostosis. The male-

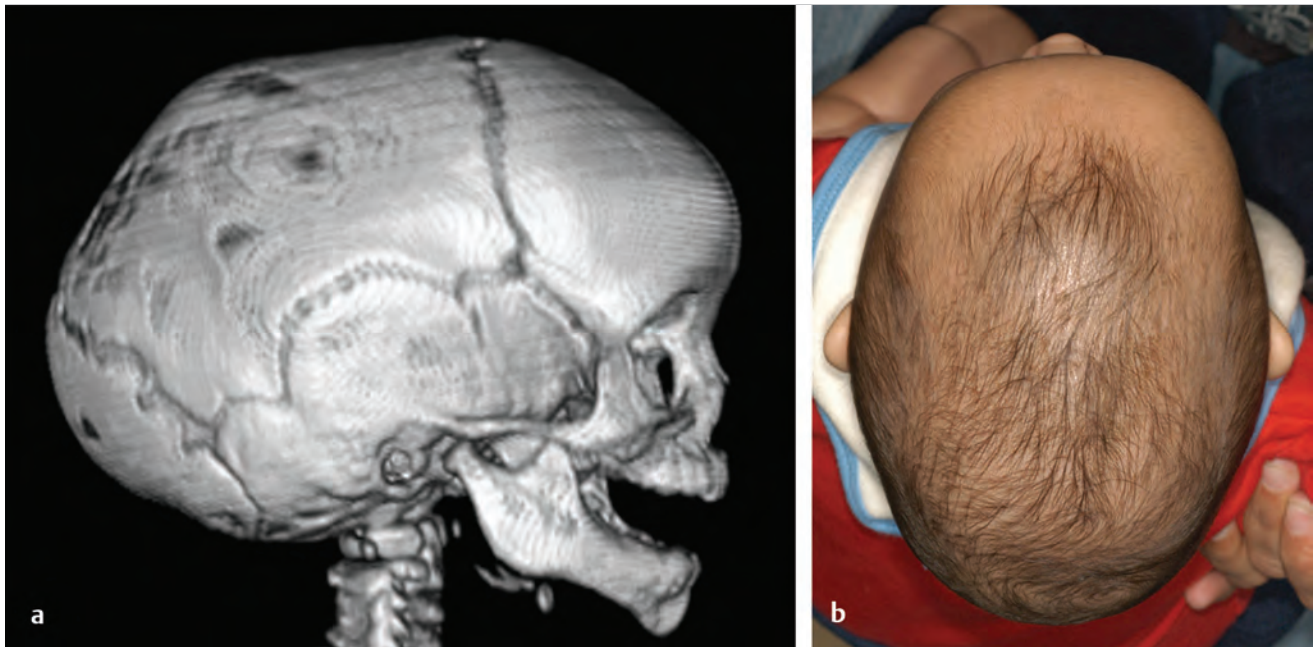


Fig. 1.7 Sagittal synostosis. (a) Lateral CT image showing cranial elongation. (b) Apical photograph of an affected patient.

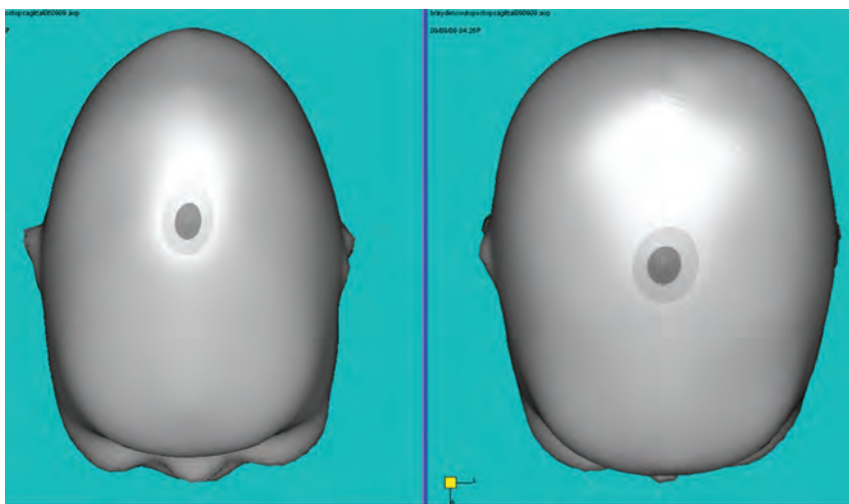


Fig. 1.8 Laser scan images demonstrating correction of sagittal synostosis by using EAS + HT.

to-female incidence ratio is 4:1. Isolated sagittal synostosis is rarely associated with a syndrome or genetic mutation, but familial examples have been reported. The characteristic phenotype consists of an elongated anteroposterior dimension and a decrease in biparietal width (► Fig. 1.7). There is often a ridge over the fused sagittal suture and, occasionally, a prominence over the anterior fontanelle. The long, thin head shape is termed scaphocephaly, or “boat-like.” Head circumference is typically larger than normal.

Management varies widely among institutions, but the authors prefer minimally invasive options (EAS + HT and spring-mediated distraction) in infants aged 5 months or younger. These techniques provide excellent, lasting results with minimal morbidity (► Fig. 1.8). Distraction is a recent addition to the surgical armamentarium and has its advocates. It is technically similar to springs but the distractors are higher profile and

protrude through the skin. Reported complications are higher with this method, and there are no real advantages over other minimally invasive techniques. Many neurosurgeons still perform the Pi procedure (named after its semblance to the Greek letter π) in affected infants who are younger than 6 months. This open operation is midway between a remodeling and a suturectomy. It involves excision of bony segments on either side of the sagittal suture (which comprises the vertical limbs of the π) and a transverse segment of bone parallel to the coronal suture (which comprises the horizontal limb of the π) from squamosal suture to squamosal suture. Barrel stave osteotomies can be added as necessary to allow temporal/parietal out-fracturing. The anteroposterior dimension of the cranium can be reduced by advancing the sagittal strip to the frontal bone and securing it with suture or resorbable plates. Aggressive shorting can compress the brain and is not recommended.

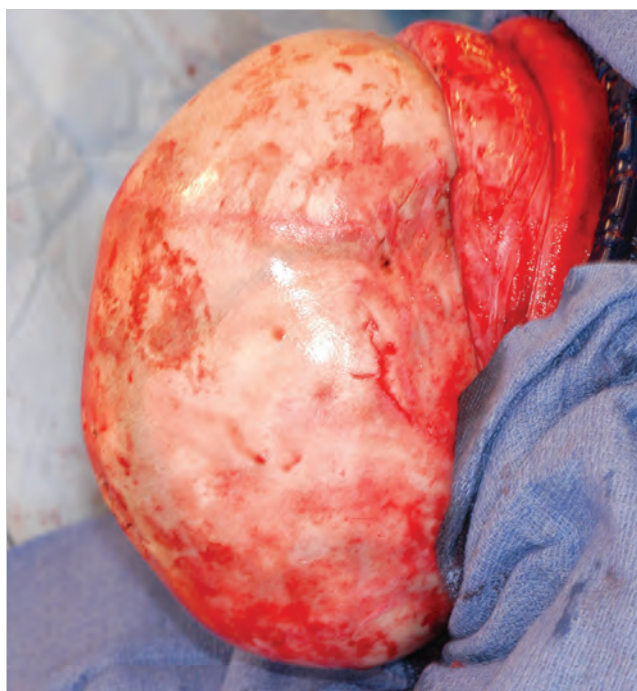


Fig. 1.9 Operative exposure of the entire cranium in the supine position.

Conventional management of patients older than 6 months is subtotal or total calvarial remodeling. In an older child, the degree of anteroposterior shortening that is safe is considerably less than that in a younger infant. Total vault remodeling for sagittal synostosis requires exposure from the glabella anteriorly to the posterior cranial base. This can be achieved by using a modified prone position, but supine positioning on a cerebellar head holder provides excellent access, allows better blending of the anterior and posterior repair, and is less risky (► Fig. 1.9). The supine position for correction of sagittal synostosis requires some head manipulation to access the posterior cranium, and it is wise to secure the endotracheal tube to the mandible or dentition before prepping. An awl can be used to pass a 26-gauge wire around the mandible; for patients with mature dentition, the wire can be affixed to the teeth. The parietal segments are removed and particulate bone is harvested from the endocortex and stored. The bones are remodeled with the goal of increasing the lateral convexity; this can be achieved with radiating cuts of increasing width peripherally. The edges are affixed with resorbable plates on the endocortical side. Low temporal and parietal regions' barrel stave osteotomies are performed, and the segments are out-fractured. This greatly expands the parietal width and provides a more complete release. The cone-shaped occiput and frontal region can be removed or, in milder cases, remodeled in situ with radial osteotomies and out-fracturing to lessen the convexity. Shortening of the anteroposterior length is not always required, but if needed, this can be accomplished by removing either the frontal or occipital bone, contouring, and replacing. We rarely reduce the AP dimension, especially in the frontal region, as this can compress the brain and may not be required. At least one study has demonstrated that frontal bossing in patients with

sagittal synostosis improves with age. Once the bone fragments are remodeled satisfactorily, they are secured with absorbable plates. The parietal plates should be affixed to the temporal bone, to the intact midline sagittal strip (in an out-fractured position), and to the expanded edges of the frontal and occipital regions. Plate stabilization ensures that the expansion and shape correction achieved intraoperatively will not relapse or collapse postoperatively (► Fig. 1.10).

1.8.2 Metopic Synostosis

The metopic suture is the first cranial suture to fuse and the only one to fuse in childhood. It begins fusing at as early as 3 months of age and is complete in nearly all patients by 6 to 8 months of age. Unlike other cranial sutures, radiographic evidence of a fused metopic suture in infancy or early childhood is not per se abnormal. Instead, it is the phenotype (i.e., the extent of forehead and superior orbital narrowing) that defines whether a radiographically closed metopic suture is considered craniosynostosis (premature and abnormal fusion) or represents normal physiologic closure. There is wide variation in the degree of forehead deformity, and the line between normal variation and "abnormal" forehead contour is poorly defined. As a consequence, there can be significant diagnostic inconsistency between centers and surgeons. On the more severe end of the spectrum, premature closure of the metopic suture results in a "keel"-shaped deformity termed trigonocephaly (► Fig. 1.11). Other findings include small, flat frontal bones; anterior displacement of the coronal sutures; and compensatory enlargement of the parietal elements. The triangular cranial shape is exaggerated by the lack of lateral projection of the supraorbital rims and narrowing of the temporal regions. Patients often have hypotelorism, strabismus, and upslanting palpebral fissures (this is sometimes referred to as the trigonocephalic "sequence").

Operative management of infants younger than 4 months includes EAS + HT. In slightly older infants (4–7 months of age), suturectomy and spring-mediated distraction provide excellent correction. Spring-mediated distraction has been shown to correct associated hypotelorism. Some centers have started using craniofacial distraction to manage this condition, although the outcomes of this are still unproven. Most craniofacial centers manage trigonocephaly with open fronto-orbital advancement. This method has predictably good outcomes and, unlike the aforementioned techniques, can be used in older infants and children. With the patient in the supine position, the frontal bones and orbital bandeau are removed. The V-shaped bandeau is expanded and, if necessary, the flat lateral segments are contoured to increase the convexity. In severe cases, the expanded bandeau may require a midline separation and leveling of each half. If this creates a gap, it can be filled with a trapezoidal or triangular bone graft. The nasal process of each frontal bone is long and should be shortened. The bandeau is intentionally overcorrected in the temporal areas and fixed with resorbable plates. The coronal gap can be filled with a rectangular piece of bone harvested from the vertex to stabilize the bandeau, provide bony continuity, and prevent any possibility of collapse. While the frontal elements are off, particulate bone graft can be harvested from the inner table and stored in a blood-filled container on the back table. This will provide bone graft to repair

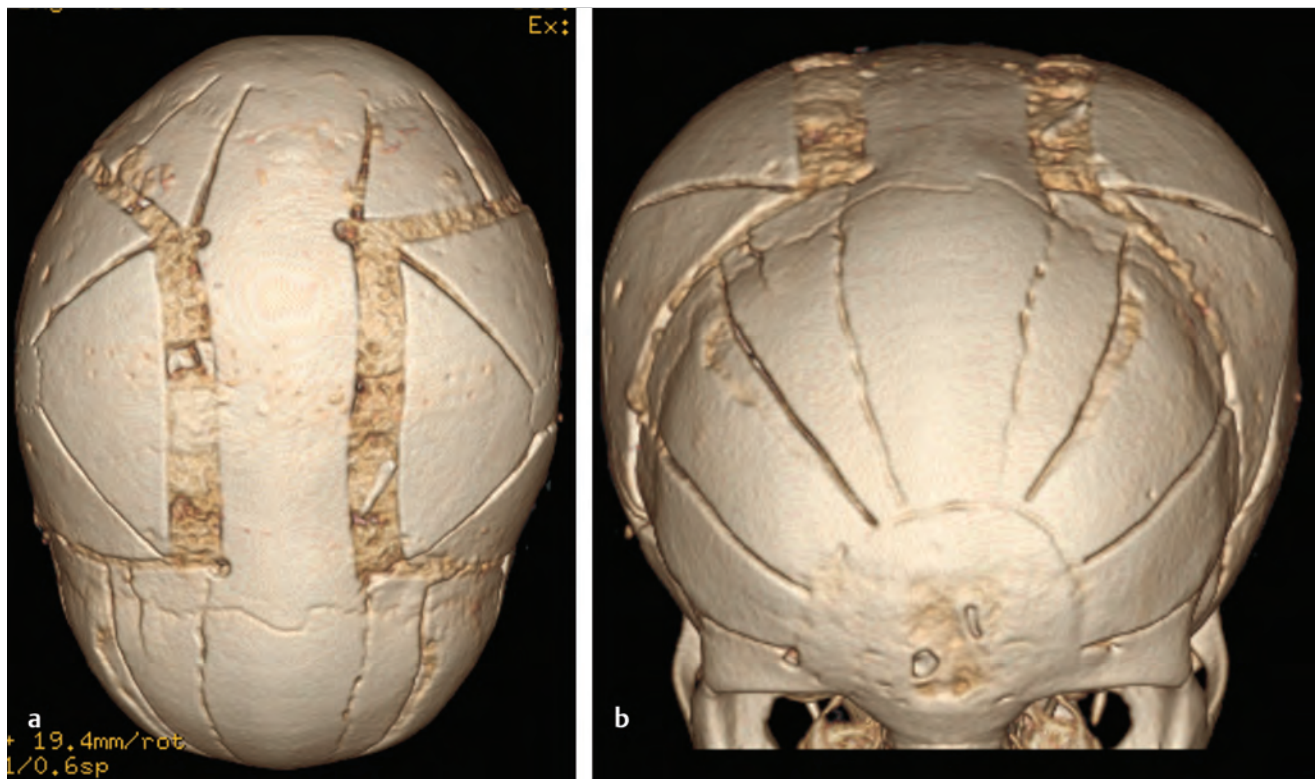


Fig. 1.10 Postoperative CT after correction of sagittal synostosis. Each parietal segment was contoured, affixed to the out-fractured temporal bone, and stabilized in a more lateral position. There is a smooth transition between the contoured regions, and all osseous defects are filled with particulate bone graft.

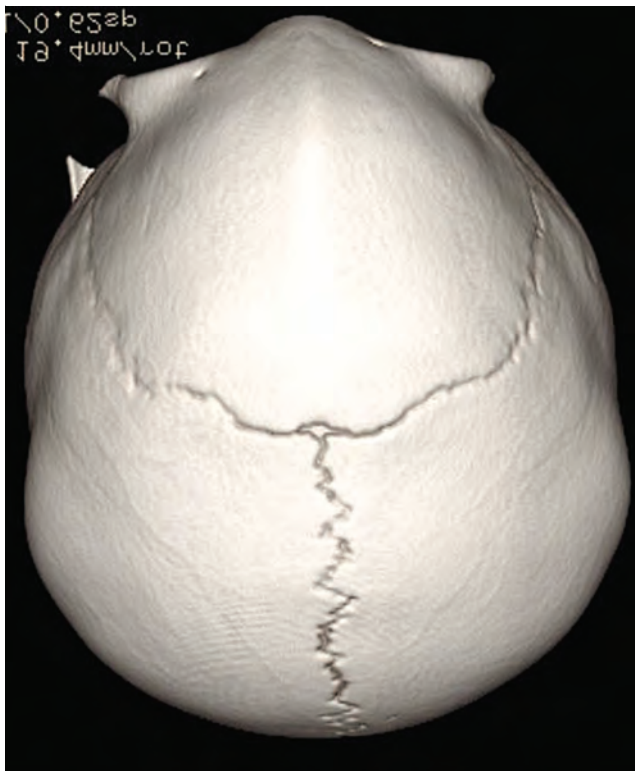


Fig. 1.11 CT image demonstrating trigonocephaly.

any defects created by the advancement or harvest of full-thickness bone graft. Each frontal bone is then remodeled with a shaping burr and/or radial osteotomies and then orthotopically or heterotopically affixed to the bandeau. The frontoparietal segments can be affixed to the bandeau with resorbable plates or secured with wires or sutures. To create a smooth transition with the widened frontal segments, the parietal bones should be laterally out-fractured by using horizontal barrel staves and stabilized to the frontal elements with absorbable plates. Although the lateral bone gaps between the frontal and parietal segments could be left to heal spontaneously, repair with full-thickness cortical bone harvested from the vertex provides bone continuity and reduces the likelihood of a depression (temporal hollow) in this area. Any bony gaps on the vertex can be filled with particulate graft stabilized with fibrin glue. The temporalis muscle should be advanced anteriorly and resuspended (► Fig. 1.12). If the soft tissue closure is tight, the galea can be scored to reduce tension on the repair. This should not induce alopecia if done properly—the scoring must not enter the fat layer where the hair follicles reside.

1.8.3 Unilateral Coronal Synostosis

Premature fusion of one coronal suture occurs in approximately 1 in 10,000 live births. This fusion results in shortening of the anterior cranial fossa; angulation of the cranial base, with shortening of the lateral cranium on the side ipsilateral to the fusion; and elevation of the greater and lesser sphenoid wings on the

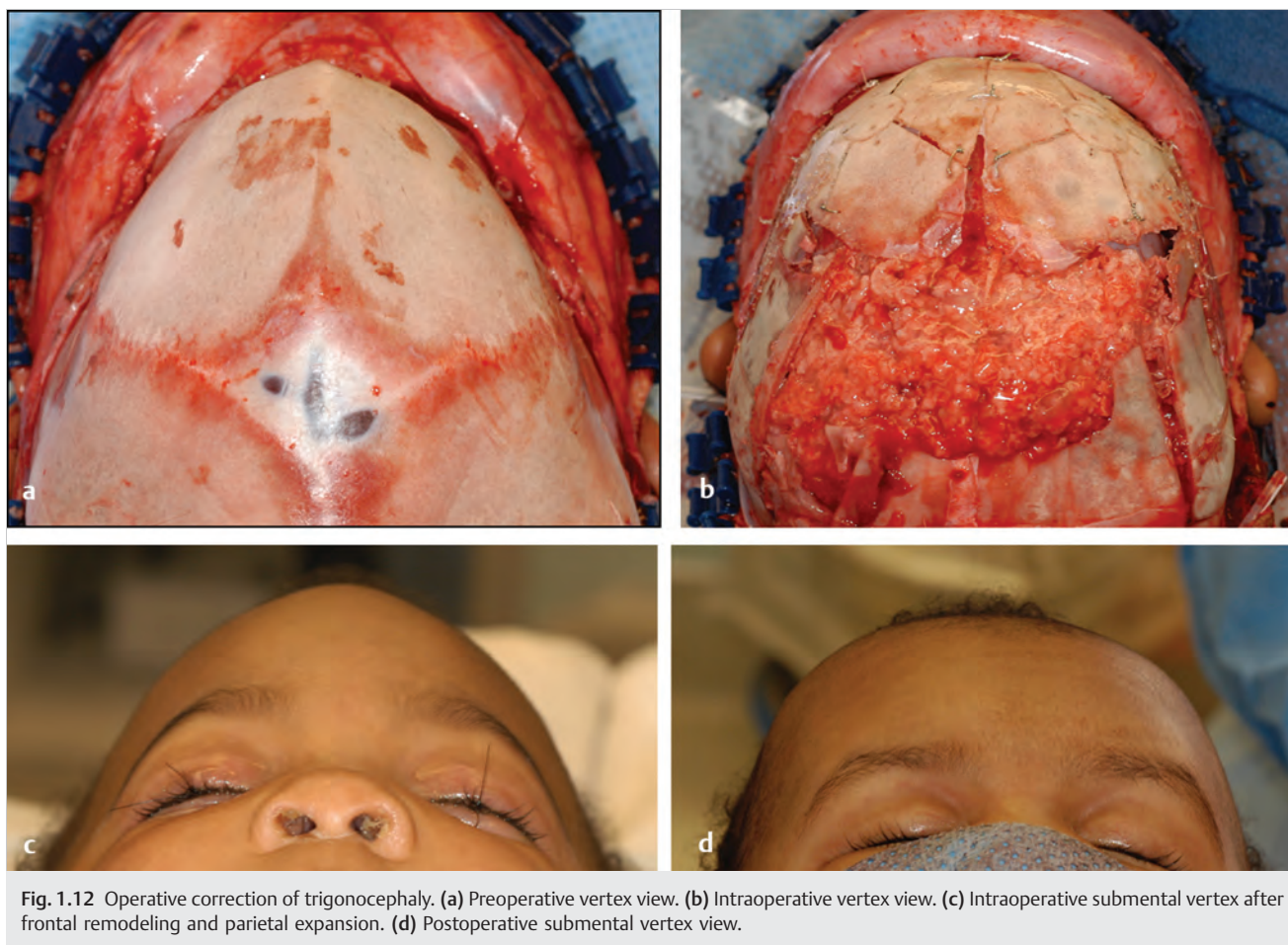


Fig. 1.12 Operative correction of trigonocephaly. (a) Preoperative vertex view. (b) Intraoperative vertex view. (c) Intraoperative submental vertex after frontal remodeling and parietal expansion. (d) Postoperative submental vertex view.

fused side leads to the characteristic “harlequin” orbit seen on a plain radiograph. There is a compensatory bulge in the ipsilateral squamous portion of the temporal bone, contralateral frontal and parietal bones, and, to a much lesser degree, the contralateral occipital bone. The fused coronal suture may demonstrate prominent ridging, and the ipsilateral frontal bone is flat. There is facial asymmetry and angulation that is pathognomonic (► Fig. 1.13). Specific findings are shortening of the ipsilateral palpebral fissure, superior and posterior displacement of the upper orbital rim and eyebrow, anterior displacement of the malar eminence, and deviation of the nasal root toward the flat frontal bone. The chin point deviates to the contralateral side. As noted earlier, the orbital changes cause ocular torticollis in approximately 80% of patients and more than half of patients have astigmatism in the eye on the unfused side. Twenty-five percent of patients have an associated molecular/syndromic diagnosis, the most common being *FGFR-3 Pro250Arg* (Muenke’s syndrome), followed by *FGFR-2*, *TWIST* (Saethre–Chotzen syndrome), and *EFNB1* (craniofrontonasal malformation).

Treatment alternatives include endoscopic suturectomy and postoperative helmet therapy as well as spring-mediated distraction for early correction of unilateral coronal synostosis. One study found a lower incidence of ocular torticollis and astigmatism in patients undergoing early endoscopic suturectomy compared with those who had later fronto-orbital advancement. In addition, early intervention may lead to better

overall facial symmetry. Most centers still treat coronal craniosynostosis by using conventional fronto-orbital advancement. After removal of the frontal bones and bandeau, each is remodeled. The frontal bone is remodeled by using radial osteotomies, and/or selective fractures are performed to achieve the desired form. The bandeau is contoured to increase convexity of the flattened side. A resorbable plate placed on the endocortical surface is excellent for maintaining the bandeau shape. There is a tendency for the correction to relapse, so many surgeons advocate recontouring and sagittal overcorrection of the bandeau and frontal bone on the side of the fusion. Some authors have recommended using an onlay bone graft over the orbital rim on the ipsilateral side to help accentuate the sagittal projection, but it is the authors’ experience that onlay grafts resorb. The bandeau is repositioned with ipsilateral over-advancement and affixed to each temporal fossa with resorbable plates. A full-thickness rectangular bone graft behind the bandeau will help prevent postoperative collapse and ensure bony healing. The frontal bone plates are reattached to the orbital bandeau with sutures or resorbable plates. The frontal bones should be attached to the parietal segments with absorbable plates to prevent collapse during redraping of the coronal flaps. Full-thickness bone grafts harvested from the vertex can be used to fill the lateral coronal gap, especially on the more advanced side. Particulate cranial bone graft harvested from the endocortex of the frontal elements, or ectocortex of the parietal bones, can

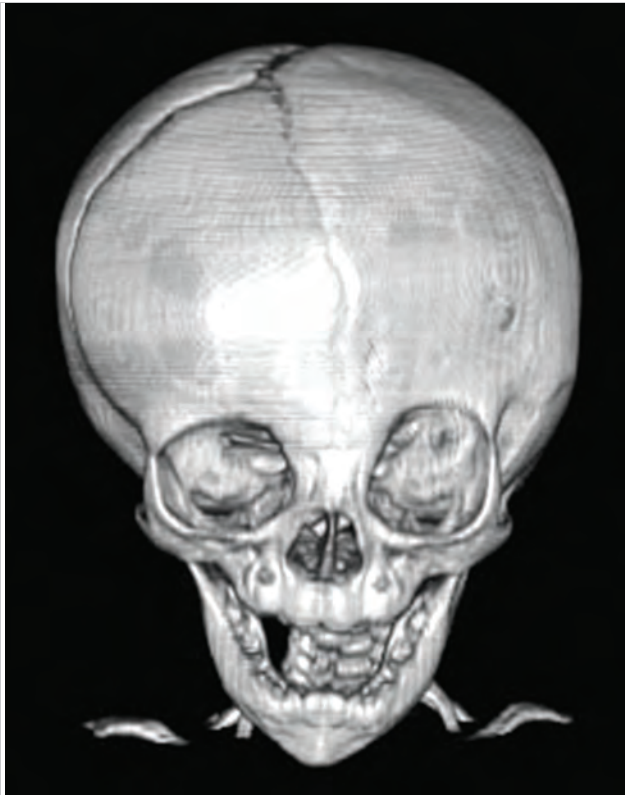


Fig. 1.13 CT image demonstrating unilateral coronal synostosis.

easily cover all remaining bone defects. The asymmetry of the palpebral fissure can be corrected with a lateral canthopexy on the side of the fusion (► Fig. 1.14).

1.8.4 Lambdoid Synostosis

Lambdoid synostosis is the least common form of craniosynostosis (1–5%) and is characterized by occipital flattening, mastoid bossing, and posterior/inferior displacement of the ear on the side of the fusion. In addition, there is decreased height of the cranial vertex on the affected side. CT scan demonstrates bony bridging of the lambdoid suture and angulation of the posterior cranial fossa toward the side of the fusion (► Fig. 1.15). The decision to recommend an operative repair for unilateral lambdoid synostosis depends on the severity of the deformity. The treatment varies depending on whether the condition is unilateral or bilateral and on the age at presentation. Minimally invasive alternatives are excellent for this condition in young infants (<7 months of age). Distraction has been advocated by some and appears to provide excellent expansion of the occiput. This technique can be used in children who present at an older age. Most centers still use open remodeling, especially in older children. Positioning is prone, so that the occipital bone can be fully visualized to the level of the foramen magnum. A bilateral parietooccipital bone segment is elevated, or each parieto-occipital bone graft can be elevated, leaving a strip of bone over the sagittal suture. Barrel stave osteotomies are performed bilaterally in the flattened basal occipital bone to increase the



Fig. 1.14 Operative correction of right unilateral coronal synostosis. (a) Preoperative submental vertex view. (b) Intraoperative oblique showing correction after fronto-orbital remodeling. (c) Intraoperative apical view of correction. (d) Postoperative submental vertex view.

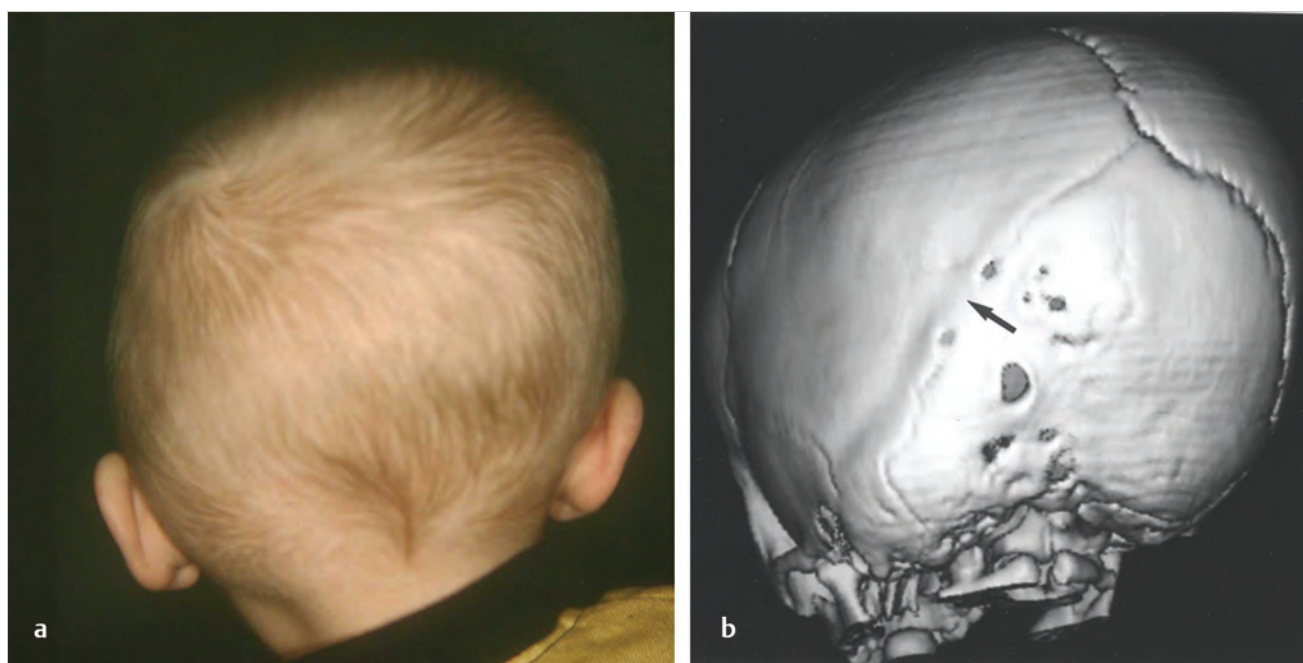


Fig. 1.15 Left lambdoid synostosis. (a) Clinical appearance. (b) CT image demonstrating fusion of left lambdoid suture.

convex projection of the occipital bone locally. The bone grafts may be orthotopically or heterotopically (switched) repositioned and secured with resorbable plates.

1.8.5 Bilateral Coronal Synostosis

Most patients (~75%) with bilateral coronal synostosis have an associated syndromic diagnosis, the most common being Crouzon's syndrome (*FGFR-2*), Muenke's syndrome (*FGFR-3 Pro250Arg*), Apert's syndrome (*FGFR-2*), Saethre–Chotzen syndrome (*TWIST*), Pfeiffer's syndrome (*FGFR-1* or *FGFR-2*), and craniofrontonasal syndrome (*EFNB1*). Thus, most infants with bilateral coronal synostosis should be presumed to have a syndromic diagnosis and should undergo genetic testing. In many instances, the syndromic diagnosis is easily ascertained by the constellation of associated physical findings. For example, Apert's syndrome is easily identified by the presence of complex syndactyly of the hands and feet and Pfeiffer's syndrome is identified by unusually wide thumbs and great toes. Some syndromes have subtle findings and require molecular testing to secure the diagnosis. The craniofacial phenotype depends on the syndromic diagnosis but includes retrusion of the midface and the superior orbital rim and turribrachycephaly (► Fig. 1.16).

Operative treatment of bilateral coronal synostosis has changed over the last decade. Traditionally, patients were managed with fronto-orbital advancement, often at 9 to 12 months of age. This is done similar to the fronto-orbital procedures described previously for metopic synostosis, except that the parietal bones typically do not require repositioning. One problem with this approach was that the combination of anterior synostosis and posterior deformational flattening from lying supine in the first 4 to 6 months of life often led to severe compensational turribrachycephaly that was difficult to reverse with an isolated anterior procedure. Recent studies have begun to emphasize on early posterior expansion, often using

distraction, and a later-staged anterior procedure to improve frontal shape. Advocates suggest that this approach provides the neurocognitive benefits of early decompression, avoids the development of compensational turribrachycephaly, and permits a later anterior reconstruction (presumably less relapse). The disadvantage of this algorithm is that it requires three procedures early in life: a relatively large procedure to create the posterior osteotomies and place the distractors, another minor procedure to remove the distractors, and the frontal procedure at some time in the future. The procedure also results in closure of the normal lambdoid sutures; the effect of this is unknown. Minimally invasive procedures have also been used to provide early decompression, and these methods can similarly avoid the development of turribrachycephaly. Critics point out that distraction provides a greater degree of expansion; however, there are no data that quantify the extent of expansion required to adequately decompress the cerebrum and offset secondary cranial changes.

Midfacial retrusion is a common feature in almost all patients with bilateral coronal synostosis and often requires operative intervention at some point during development to improve facial aesthetics and treat obstructive sleep apnea, a common finding in this patient population. The severity of midfacial retrusion varies with the associated syndrome: most severe in infants with Apert's and Pfeiffer's syndromes, moderate in patients with Crouzon's syndrome, and very mild in patients with Muenke's and Saethre–Chotzen syndromes. The timing of midfacial advancement is, consequently, largely determined by the type of associated syndrome.

1.8.6 Unusual Craniosynostosis Variants

Although the majority of patient with craniosynostosis fall within one of the categories listed earlier, there are many

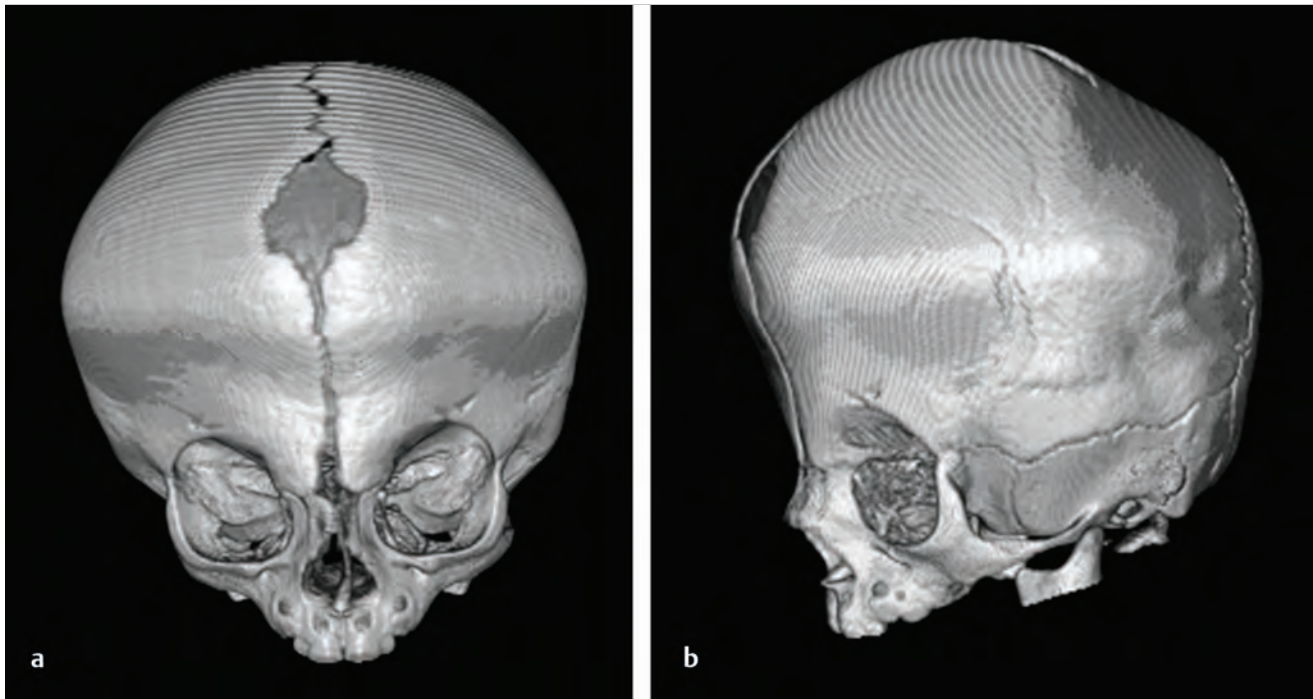


Fig. 1.16 CT images of bilateral coronal synostosis. (a) Anteroposterior view. (b) Lateral view.

unusual and rare variations that are worth examining. Most of these create such an unusual cranial shape that they are noticed by even most inexperienced clinician, but unfortunately, definitive diagnosis can be delayed by the presumption that the shape is a product of cephalic molding from the birthing process. For example, the combination of metopic and unilateral coronal synostoses yields a phenotype that is hard to dismiss as normal (► Fig. 1.17). There are sutural fusions that lead to esoteric change in cranial and facial shape and can be easily dismissed. Examples include isolated frontosphenoidal synostosis, which is phenotypically reminiscent of deformational plagiocephaly, and progressive postnatal pansynostosis (PPP; ► Fig. 1.18). Patients with the latter condition fuse all of their sutures sometime after birth and present later in childhood with a normal-appearing cranial shape, declining percentile head circumference, and elevated ICPs. Because the cranial shape is normal, the diagnosis is often delayed.

Given the rarity of these unusual forms of craniosynostosis, treatment must be tailored to the individual child. Operative treatment of these infants present a challenge to even the most experienced craniofacial surgeons, but the goals of treatment for these unusual forms of craniosynostosis are the same as those for all other types of craniosynostosis—optimize neurocognitive development and craniofacial aesthetics. Fortunately, the techniques used for more common types of fusions (described previously), in combination with the ingenuity and creativity of the craniofacial surgeon, are more than adequate to restore volume and shape in even the most severely affected patient.

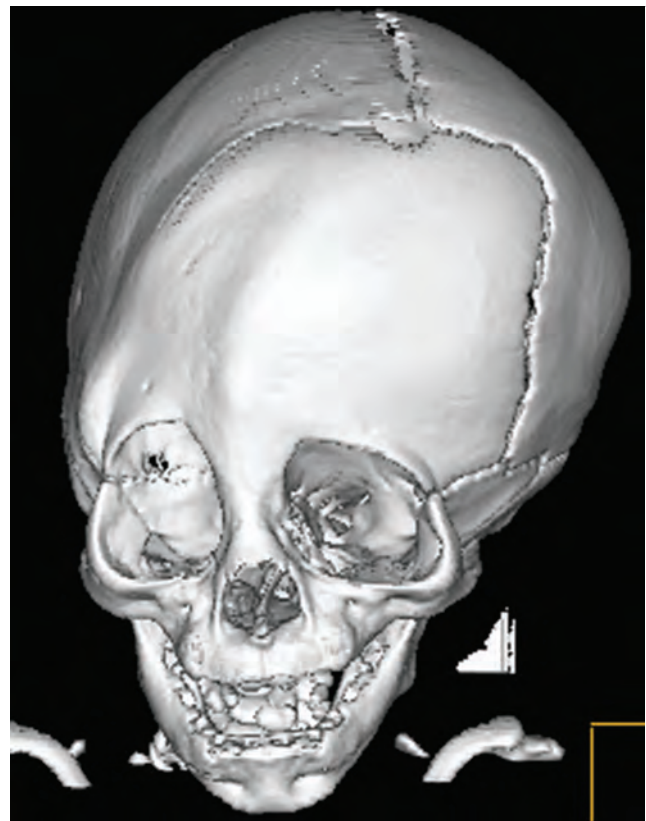


Fig. 1.17 CT image of combined right unilateral coronal and metopic synostosis.

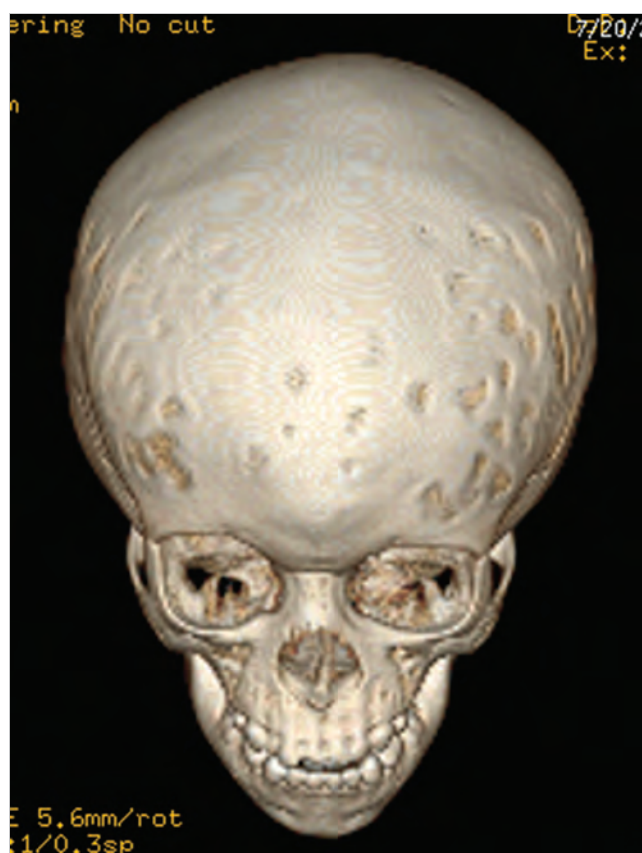


Fig. 1.18 CT image of a 2-year-old child with progressive postnatal pansynostosis. Note absence of any suture definition and surface irregularities suggestive of bone thinning.

1.9 Complications

Complications are relatively infrequent and may be divided into those that are acute and those that are late. Acute complications include major blood loss, air embolus, dural tear with cerebrospinal fluid (CSF) leak, infection, and respiratory complications. Blood loss occurs continuously during open cranial vault procedures but is most substantial during completion of the osteotomies. Bleeding is the direct or indirect cause of most complications. Consequently, attention to accurate blood replacement is paramount to avoid coagulopathy secondary to dilution of clotting agents. It is critical to have sufficient IV access to allow rapid resuscitation. Blood loss may continue for 12 to 24 hours following large cranial remodeling procedures, and intensive care unit monitoring is usually warranted. Air embolus has been documented in children undergoing cranial procedures and may occur in any operative position, including supine. The placement of precordial Doppler monitors and end-tidal CO₂ monitors to ascertain entrainment of air into the

venous system is recommended. Central venous lines are sometimes warranted to assess blood volume (and resuscitation) and can be helpful to evacuate an air embolus if it occurs.

Dural tears are not uncommon and should be identified and repaired. An unrepaired or incompletely repaired dural tear can result in a persistent CSF leak into the drain (if one is used) or an unresolved fluid collection under the closed coronal flaps. Persistent CSF leak can lead to infection, thinning of the overlying bone, and a cranial defect. Infection is an uncommon problem after cranial procedures but can be potentially life-threatening. Infection can arise from a persistent CSF leak or a communication of the intracranial cavity with the nasal cavity or frontal sinus. Because the frontal sinus develops quite late in childhood, the latter is seen as a consequence of surgery in older children.

Late complications include incomplete bone healing, impaired bone growth, relapse, and recrudescence of the original cranial deformity. Several studies have reported a negative correlation between age at repair and recurrence. The degree of relapse may also depend on the severity of the initial phenotype as well as the continued effect of cranial base restriction. Other important factors may be incomplete correction and inadequate bone stabilization. Overall, the morbidity and mortality resulting from the treatment of craniosynostosis are quite low. Mortality has been variously reported to range between 1.5 and 2%. Current advances in monitoring and anesthetic techniques, as well as refinements in surgical techniques, have driven this rate well below 1% at most large centers.

Suggested Readings

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2 Craniofacial Syndromes

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Summary

The diagnosis and management of syndromic craniofacial anomalies, namely Apert, Crouzon, Muenke, Pfeiffer, Saethre-Chotzen, and Treacher Collins syndromes, are complex due to the wide phenotypic variability of the clinical presentations. Craniofacial syndromes can be divided into two major categories: craniosynostosis-associated syndromes and cleft-associated syndromes. Common syndromic craniosynostosis include Apert, Crouzon, Muenke, Pfeiffer, and Saethre-Chotzen, while Treacher Collins is the most common cleft-associated mandibulofacial dysostosis condition. Management of patients with craniofacial syndromes includes correctly timed surgical operations in a staged fashion, close involvement of the patients' families, as well as psychosocial and aesthetic considerations for the pediatric patients. A multidisciplinary team approach is vital to a successful outcome, and the involvement of a pediatric plastic surgeon in the surgical management of these complex craniofacial syndromes is central.

Keywords: Craniofacial syndromes, congenital disorders, genetic mutations, Apert syndrome, Crouzon syndrome, Muenke syndrome, Pfeiffer syndrome, Saethre-Chotzen syndrome, Treacher Collins syndrome

2.1 Introduction

Craniofacial syndromes can be divided into two broad categories: craniosynostosis-associated syndromes and cleft-associated syndromes. The clinical diagnosis and surgical management of craniofacial syndromes, such as Apert's, Crouzon's, Muenke's, Pfeiffer's, Saethre-Chotzen, and Treacher Collins syndromes, are

complex due to the overlapping *and* varied phenotypic presentations and genetic mutations of the different syndromes. For example, all craniosynostosis syndromes have abnormal, premature fusion of one or more cranial sutures. However, some syndromes, such as Apert's and Pfeiffer's syndromes, have prominent midface extrusion, whereas the midface position is less severely affected in other conditions, such as Muenke's and Saethre-Chotzen syndromes. Thus, correct diagnosis and treatment for these conditions require a multidisciplinary team approach. Surgical and nonsurgical management needs to be tailored based not only on the genetic and clinical diagnosis but also on the severity of symptoms and functional compromise experienced by the patient. Optimal treatment quite often necessitates multiple surgical disciplines, with the involvement of a pediatric plastic surgeon being essential for a successful outcome.

Craniosynostosis results from the premature fusion of one or more cranial sutures and is a relatively common congenital defect, affecting 1 in every 2,000 births globally. Premature fusion restricts the growth of the skull perpendicular to the suture involved; consequently, compensatory skull growth occurs parallel to the affected suture, in order to accommodate the growing brain (► Fig. 2.1). Craniosynostosis usually occurs as an isolated condition but can also result from a detectable genetic mutation. Apert's, Crouzon's, Muenke's, Pfeiffer's, and Saethre-Chotzen syndromes are unique for their designation as syndromic craniosynostoses and are collectively the most common syndromic craniosynostoses. More than 100 syndromes have been described and attributed to specific genetic mutations, notably *fibroblast growth factor receptor-1* (*FGFR-1*), *FGFR-2*, and *FGFR-3*, and the transcription factors *TWIST* (upstream regulator of *FGFRs*) and *MSX-2*. Specifically, gain-of-

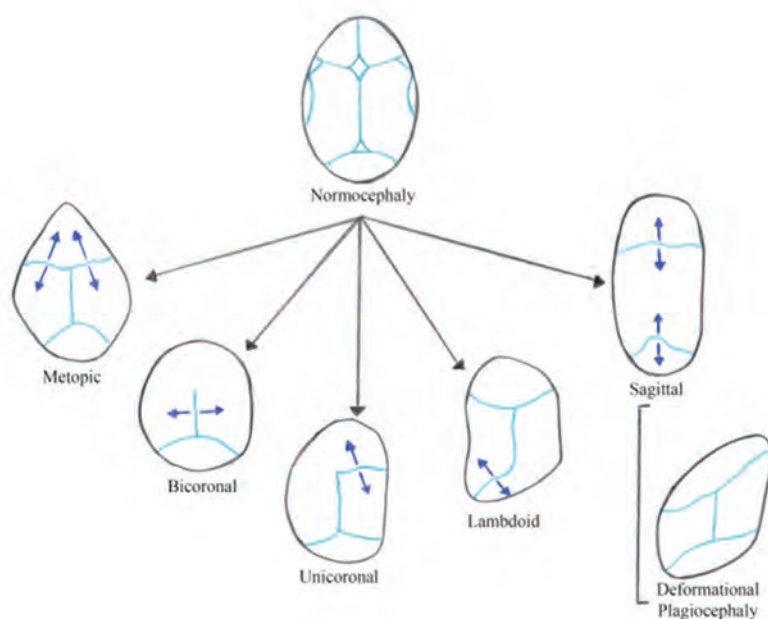


Fig. 2.1 Schematic of craniosynostosis deforming forces.

Table 2.1 Overview summary of craniofacial syndromes

Syndrome	Epidemiology	Category	Causative gene	Inheritance	Clinical features
Apert's	1 in 65,000 live births	Craniosynostosis	<i>FGFR-2</i> (10q26.13)	Autosomal dominant	Craniosynostosis (usually bicoronal), midface hypoplasia, shallow orbits, beaked nose, conductive hearing loss, complex acrosyndactyly of hands and feet
Crouzon's	1 in 60,000 live births	Craniosynostosis	<i>FGFR-2</i> (10q26.13)	Autosomal dominant	Craniosynostosis (usually bicoronal), midface hypoplasia, shallow orbits, hearing loss
Muenke's	1 in 30,000 live births	Craniosynostosis	<i>FGFR-3</i> Pro-250-Arg mutation (4p16.3)	Autosomal dominant	Craniosynostosis (bicoronal or unicoronal), midface hypoplasia with ocular hypertelorism, hearing loss, variable and low penetrance limb malformations
Pfeiffer's	1 in 100,000 live births	Craniosynostosis	<i>FGFR-1</i> (8p11.23-p11.22), <i>FGFR-2</i> (10q26.13)	Autosomal dominant	Craniosynostosis (usually bicoronal), midface hypoplasia, shallow orbits, hearing loss, broad radially deviated thumbs, widened great toes, partial syndactyly of fingers and toes
Saethre–Chotzen	1 in 25,000 to 1 in 50,000 live births	Craniosynostosis	<i>TWIST1</i> (7p21.1)	Autosomal dominant	Craniosynostosis (usually bicoronal), syndactyly, congenital heart defects, low frontal hairline, ptosis, small ears with prominent horizontal crura
Treacher Collins	1 in 50,000 live births	Cleft	<i>TCOF1</i> , <i>POLR1D</i> (5q31.3-q33.3) 5 (5q31.3-q33.3), or autosomal recessive for <i>POLR1C</i> (6p22.3)	Autosomal dominant (<i>TCOF1</i> , <i>POLR1D</i>) Rarely, autosomal recessive (<i>POLR1C</i>)	Cleft palate, malar and mandibular hypoplasia, downward-slanting palpebral fissures, coloboma of the lower lid, microtia, conductive hearing loss

Source: Upton 1991, Grieg and Dunaway 2015.

function mutations are associated with the *FGFR* genes and human *MSX-2*, whereas loss-of-function mutations are associated with the *TWIST* gene. Mutations in *FGFR-2* gene are the most prevalent cause for three notable, overlapping syndromes: Apert's, Crouzon's, and Pfeiffer's syndromes (► Table 2.1).

Surgical intervention remains the only treatment to correct the underlying pathology. The main goals of surgery are expansion and normalization of the calvarial shape to increase the intracranial volume and normalization of craniofacial proportions and occlusal relationships. Early (< 1 year of age) procedures aim to allow adequate intracranial volume for brain growth and avoid potential consequences of increased intracranial pressure. Surgical procedures during childhood and teenage years aim to correct midface retrusion and to improve the airway, bony orbital volume, and occlusion. The two main surgical procedures for craniosynostosis repair are fronto-orbital advancement (FOA) with open cranial vault remodeling and

strip craniectomy, which can be performed in a minimally invasive endoscopic-assisted fashion. Each approach has its own advantages and disadvantages and may or may not be indicated based on the type of craniosynostosis and the age of presentation. Complications associated with surgical correction include bleeding, infection, optic nerve ischemia, seizures, delayed healing and other wound problems, incomplete bone healing resulting in chronic bone defects, contour irregularities, and need for additional surgical procedures.

Treacher Collins syndrome differs from the aforementioned syndromes in that it is not a syndromic craniosynostosis. By contrast, Treacher Collins syndrome is the most common cleft-associated syndrome and is also known as mandibulofacial dysostosis. Occurring in 1 in 50,000 live births, Treacher Collins syndrome is an autosomal dominant disorder of craniofacial development, and its major clinical features include bilateral absence or hypoplasia of the zygomas, cleft palate, midface

hypoplasia, micrognathia, microtia, and conductive hearing loss. Recently, mutations in *TCOF1* (Treacher Collins–Franceschetti syndrome 1) and *POLR1D*, in chromosome 5, have been identified as the cause of Treacher Collins syndrome. The goals of surgical treatment and overall management of patients with Treacher Collins syndrome include palate repair, speech therapy, construction of ears, reconstruction of the zygomatic prominence, and correction of midface retrusion to improve the airway and respiratory status as well as to improve occlusion. Treacher Collins syndrome highlights and provides an excellent example of the complex, surgically staged, multidisciplinary team approach requisite for the care of patients with congenital disorders, particularly those with cleft palate.

Herein, we describe the clinical diagnosis and management of six syndromes, Apert's, Crouzon's, Muenke's, Pfeiffer's, Saethre–Chotzen, and Treacher Collins syndromes, which together are reflective of the wide spectrum of craniofacial syndromes confronting pediatric plastic and reconstructive surgeons today.

2.2 Diagnosis

2.2.1 Craniosynostosis-Associated Syndromes

Apert's syndrome, or acrocephalosyndactyly type I, is seen in 1 in 65,000 live births and accounts for 4.5% of all craniosynostosis cases. Characteristic clinical features of Apert's syndrome are craniosynostosis (usually bicoronal), turribrachycephaly, down-slanting palpebral fissures, hypertelorism, exorbitism, cleft palate, midface hypoplasia or retrusion, and complex syndactyly of the hands and feet. The syndrome is caused by one of the two *FGFR-2* mutations on chromosome 10 (10q26.13), Ser-252 and Pro-253. It either is inherited in an autosomal dominant fashion or can arise *de novo*, with older paternal age being a recognized risk factor for the *de novo* mutation. A specific missense substitution involving adjacent amino acids, Ser-252-Trp and Pro-253-Arg, in the linker between the second and third extracellular immunoglobulin domains of *FGFR-2* has been identified as the causal mutation resulting in Apert's

syndrome. Interestingly, there is a varying genotype–phenotype correlation among the two noted *FGFR-2* mutations, Ser-252 and Pro-253. Cleft palate, nasolacrimal stenosis, and severe ocular findings, such as ptosis, strabismus, and amblyopia, are more common in patients with the Ser-252 mutation in *FGFR-2*. However, the degree of syndactyly and intellectual disability is more significant in patients with the Pro-253 mutation in *FGFR-2*. Moreover, males and females are affected equally, and 50% of children with a parent diagnosed with Apert's syndrome will inherit the condition. To confirm the diagnosis in a proband, targeted mutation analysis of *FGFR-2* for the mutations Ser-252-Trp and Pro-253-Arg should be conducted, and if normal, sequence analysis for rare *FGFR-2* mutations or partial gene insertions or deletions should be considered. In terms of embryological etiology, Apert's syndrome is classified as a brachial arch syndrome affecting the first brachial arch, which is the precursor to the maxilla and mandible.

The two most notable clinical features of Apert's syndrome are craniosynostosis and severe syndactyly of the hands and feet (► Fig. 2.2 and ► Fig. 2.3). The Apert's hand is a distinctive and pathognomonic feature of the syndrome and is well described by the Upton's classification (► Table 2.2). As Upton specified, the four most distinguishing features of the Apert's hand are as follows: (1) a short thumb with radial deviation, (2) complex syndactyly of the index, long, and ring finger, (3) symbrachyphalangism, and (4) simple syndactyly of the fourth web space. In fact, Apert's syndrome is most easily clinically differentiated from Crouzon's syndrome by the soft tissue and bony syndactyly in the hands and feet, which are not seen in Crouzon's syndrome. In addition, the facial and cranial features of Apert's syndrome are more prominent at birth than with Crouzon's syndrome, in which features develop progressively throughout infancy.

Other cephalometric diagnostic features of Apert's syndrome include a shortened and widened anterior cranial fossa and reduced bony orbital volume, leading to exorbitism. In the mid-sagittal plane, there is a wide-open calvarial defect from the root of the nose to the posterior fontanelle. Cloverleaf skull, or kleeblattschadel, has been rarely reported in association with Apert's syndrome. There is a high incidence of cleft palate at



Fig. 2.2 Associated non-craniofacial findings are seen in certain craniosynostosis syndromes. Simple and complex syndactylies are associated with Apert's syndrome (photographs courtesy of Dr. Jeffrey A. Ascherman, Columbia University Medical Center, New York, NY).

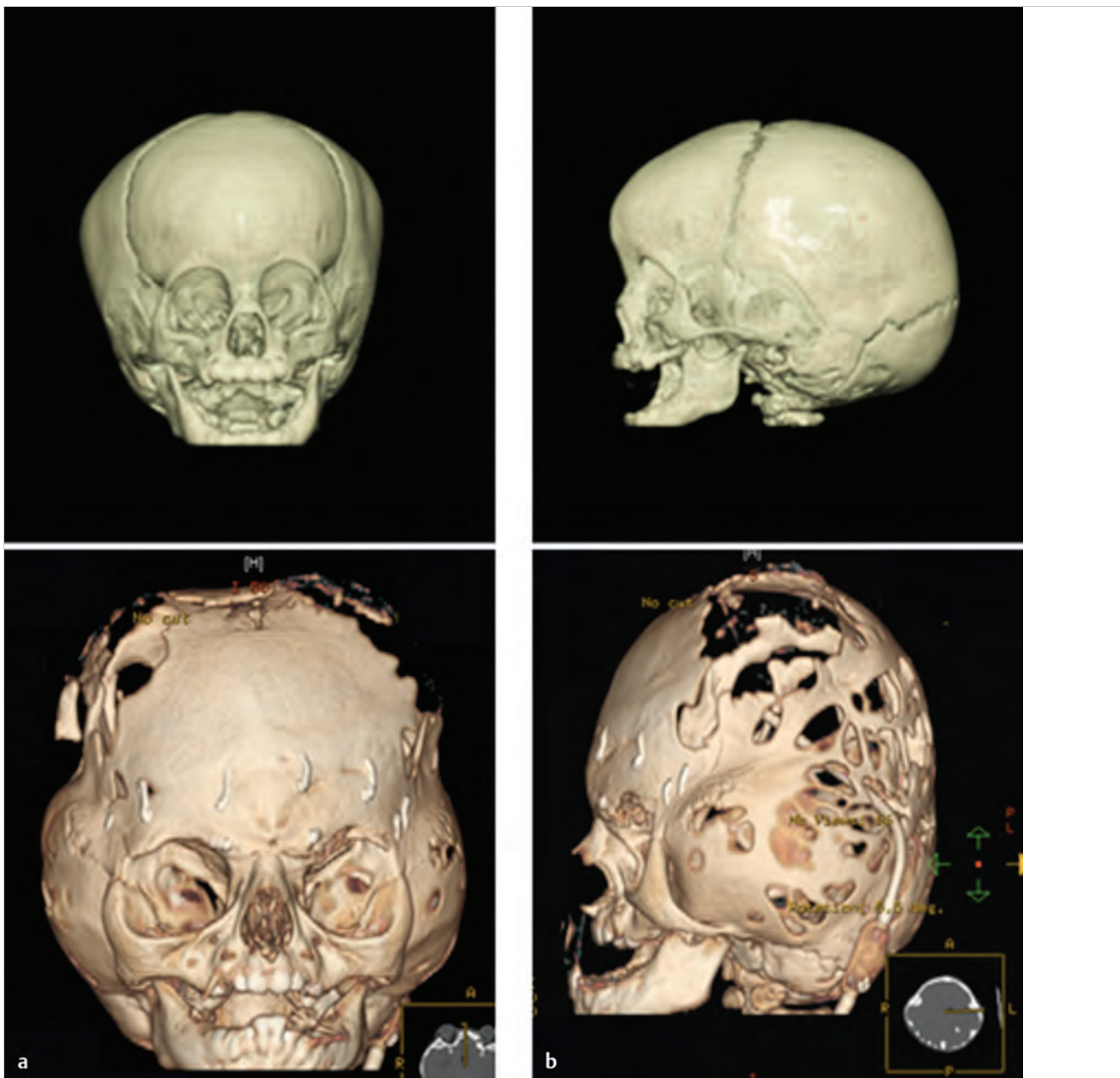


Fig. 2.3 Three-dimensional CT scan demonstrates the bilateral coronal synostosis and midface retrusion common in many syndromic craniosynostosis cases, as compared to nonsyndromic, single-suture synostosis. **(a)** Metopic synostosis with patent coronal sutures and no midface retrusion. **(b)** Apert's syndrome with bilateral craniosynostosis and midface retrusion.

50%, divided equally with 25% true clefts and 2% pseudo-clefts. The majority of patients with Apert's syndrome have a high-arched palate, which is seen in conjunction with an anterior open bite and dental overcrowding. Notably, there is no association with cleft lip. The lips are crossbow-shaped or trapezoidal, and the lower lip usually protrudes. Ocular findings include possible globe subluxation, downward slant of the lateral canthi, and a well-recognized S-shaped upper-eyelid ptosis. Finally, there is often significant cutaneous involvement, with abnormally thick skin encapsulating skeletal abnormalities, seborrhea, and eventually facial acne as the patient ages.

Sequelae of Apert's syndrome include, but are not limited to, poor intellectual development, obstructive sleep apnea (OSA),

repeated ear or sinus infections, hearing loss, and ophthalmic morbidity, namely strabismus with more esotropia than exotropia.

Crouzon's syndrome, or craniofacial dysostosis type I, has an incidence of 1 in 60,000 live births in the United States (1 in 25,000 globally) and accounts for 4.8% of all craniosynostosis cases. The syndrome is caused by a mutation in *FGFR-2*, and the specific gene is on chromosome 10 (10q26.13). A specific mutation within the *FGFR-3* gene accounts for Crouzon's syndrome and acanthosis nigricans. Similar to Apert's syndrome, Crouzon's syndrome is an autosomal dominant genetic disorder as well as a syndrome affecting the first brachial arch, which is the precursor for the maxilla and mandible. A tall, flattened

Table 2.2 Upton's classification system of the Apert's syndrome hand (1991)

Deformity	Type I "Spade hand" (most common)	Type II "Mitten hand"	Type III "Hoof or rosebud hand"
Thumb radial clinodactyly	Present	Present	Present
Index radial clinodactyly	Absent	Present	Present
First web syndactyly	Simple (nonosseous)	Simple (nonosseous)	Complex (osseous)
Complex 2–3–4 syndactyly and symbrachyphalangism	Present	Present	Present
4–5 syndactyly	Simple, incomplete	Simple, incomplete	Simple, complete

Source: Upton 1991, Grieg and Dunaway 2015.

forehead secondary to usually bicoronal craniosynostosis, a beaked nose, exorbitism, and mid-face hypoplasia characterize the syndrome. Usually, Crouzon's syndrome has a milder degree of craniofacial deformity clinically when compared with Apert's syndrome. In contrast to Apert's syndrome, patients with Crouzon's syndrome rarely have cleft palate and usually have structurally normal hands and feet as well as relatively normal intelligence.

Diagnosis of Crouzon's syndrome is most often made by clinical assessment in infancy. Clinical suspicion is heightened when the common triad of craniosynostosis, midfacial hypoplasia, and exorbitism is recognized. Further confirmatory studies include genetic testing, skull radiographs, magnetic resonance imaging scans, and computed tomographic scans. Craniosynostosis is usually bicoronal, and in many cases, the synostosis can be progressive in patients with Crouzon's syndrome. The craniosynostosis of Crouzon's syndrome is typically associated with a distinctive shortening of the anterior cranial fossa depth, with orbital roof hypoplasia and advancement of the posterior wall of the orbits, thus resulting in a severe ocular proptosis and shallow orbits. Cervical abnormalities may be part of the syndrome and can include butterfly vertebrae and fusions of posterior elements. In addition, features of Crouzon's syndrome include conductive hearing loss, strabismus, anterior overbite, short upper lip, mandibular prognathism, hypoplastic maxilla, and small nasopharynx, necessitating mouth breathing.

Muenke's syndrome has an incidence of 1 in 30,000 live births and is the most common form of syndromic craniosynostosis, accounting for up to 8% of all cases of craniosynostosis. The syndrome is autosomal dominant and is due to a mutation on the *FGFR-3* gene, specifically a Pro-250-Arg base substitution mutation that is found in 100% of the patients with the syndrome. It was the first clinical syndrome that was defined first on a molecular basis by genetic mutation.

Clinically, Muenke's syndrome presents with craniosynostosis, usually bicoronal (71% of patients) or unicoronal (29% of patients), and can occasionally present with megaloccephaly, without craniosynostosis. Patients have varying degrees of mid-face hypoplasia, with ocular hypertelorism and strabismus. In contrast to Apert's syndrome, patients with Muenke's syndrome usually have normal hand and foot phenotypes but can reportedly be found to have carpal or tarsal fusions. Unlike the hearing loss characteristic of other syndromic craniosynostosis, patients with Muenke's syndrome suffer from sensorineural hearing loss. Finally, developmental delay and intellectual disabilities have been reported in 33% of patients with Muenke's syndrome.

Pfeiffer's syndrome, or acrocephalosyndactyly type V, has an incidence of 1 in 100,000 live births and is notable among the group of syndromic craniosynostosis for its high mortality and need for multiple surgical interventions (► Fig. 2.4). Genetically, the syndrome is autosomal dominant, with incomplete penetrance and variable expressivity, and is linked to two genes *FGFR-1* (8p11.23–p11.22) and *FGFR-2* (10q26.13). However, unlike Muenke's syndrome, where there is a one-to-one genotype–phenotype correlation for the syndrome, Pfeiffer's syndrome boasts a wide spectrum of causal mutations and genotypic variability; thus, the genetic diagnosis should be approached systematically. To confirm the diagnosis of Pfeiffer's syndrome, sequence analysis of exons 8 and 10 should be done, since these are the locations for approximately 80% of *FGFR-2* mutations. If normal, then sequence analysis of exons 3, 5, 11, 14, 16, and 17 should be pursued, since these are the sites of 10% of *FGFR-2* mutations. Testing of *FGFR-1* mutation should be considered if an *FGFR-2* mutation is not found or if the patient has a milder clinical phenotype.

Clinically, broad thumbs and big toes, valgus deformities, and, occasionally, cardiac abnormalities distinguish patients with Pfeiffer's syndrome from those with other syndromic craniosynostosis. In fact, measuring the angles between extended digits of the hands can facilitate a clinical diagnosis of Pfeiffer's syndrome. In addition, synostoses of the elbow and knee, hydrocephalus, and imperforate anus are reported frequently as associated with Pfeiffer's syndrome.

Owing to the wide phenotypic and genotypic variability of the syndrome, Pfeiffer's syndrome has been further classified into three types according to a classification system described by Cohen et al (► Table 2.3). Pfeiffer's syndrome type I involves the classic, common phenotype of symmetric bicoronal craniosynostosis, variable syndactyly, broad thumbs, widened great toes, normal intelligence, and relatively normal life span (or rather survival to adulthood). Pfeiffer's syndrome type II has characteristic findings of severe ocular proptosis, ankylosis of the elbows, broad thumbs, widened great toes, severe central nervous system involvement with hydrocephalus, and visceral anomalies and carries a poor prognosis. Pfeiffer's syndrome types II and III have significantly more severe phenotypes, with bicoronal craniosynostosis in addition to multiple other sutures, as well as severe developmental delay and shortened life spans. Of note, the cloverleaf skull anomaly (kleeblattschädel) is unique to Pfeiffer's syndrome type II.

Saethre–Chotzen syndrome, or acrocephalosyndactyly type III, has an incidence in the range of 1 in 25,000 to 1 in 50,000



Fig. 2.4 (a) A nonsyndromic unilateral craniosynostosis versus (b,c) Pfeiffer's syndrome. Note the proptosis and midface retrusion in the syndromic patient.

Table 2.3 Cohen's classification system of Pfeiffer syndrome (1993)

Pfeiffer's syndrome type	Major characteristics	Associated anomalies	Prognosis
Type I	Turribrachycephaly, midface retrusion, broad thumbs, widened great toes, brachydactyly, variable syndactyly	Cervical spine fusions, low incidence of visceral anomaly	Survival to adulthood
Type II	Cloverleaf skull, severe ocular proptosis, severe mid-face retrusion, digital anomaly	Spine anomaly, elbow synostoses, intellectual delay, intracranial anomaly	Poor
Type III	Absence of cloverleaf skull, severe midface retrusion, ocular proptosis, class III malocclusion, digital anomaly	Elbow/knee synostoses, visceral anomaly, intellectual delay, intracranial anomaly	Poor

Source: Cohen 1993, Buchanan et al 2014.

live births, and it is one of the most common types of syndromic craniosynostosis. The syndrome is autosomal dominant, with high penetrance and variable expressivity, involving mutations of the *Twist1* gene on chromosome 7 (7p21.1).

Clinically, classic diagnostic findings for Saethre–Chotzen syndrome include unilateral or bilateral coronal craniosynostosis, strabismus, ptosis, low frontal hairline, and malformed ears, each with a distinctly small pinna and prominent superior crus. In contrast to Apert's syndrome, patients with Saethre–Chotzen syndrome neither have syndactyly features of the hands and feet nor have the proptosis feature characteristic of Crouzon's syndrome. Patients with Saethre–Chotzen syndrome are clinically differentiated by their low-set frontal hairline, parrot-beaked nose, brachydactyly, and milder bone deformities. In addition, patients may have partial syndactyly of the fingers, usually between the web space of the index finger and long finger.

2.2.2 Cleft-Associated Syndrome

Treacher Collins syndrome, or mandibulofacial dysostosis, has an incidence of 1 in 50,000 live births and differs from the previously discussed five craniofacial syndromes in that it is not a syndromic craniosynostosis. Treacher Collins syndrome types I and II are always autosomal dominant and are caused by one of the two genes, *TCOF1* or *POLR1D* (polymerase I, RNA, subunit D) on chromosome 5 (5q31.3–q33.3). Very rarely, Treacher Collins syndrome is autosomal recessive due to a *POLR1C* (polymerase I, RNA, subunit C) mutation on chromosome 6 (6p22.3). Although *TCOF1*, which accounts for 90 to 95% of cases, and *POLR1D* mutations are associated with extreme clinical variability, there is no specific phenotype–genotype correlation for the various signs and symptoms of Treacher Collins syndrome. In contrast to Apert's and Crouzon's syndromes, Treacher Collins



Fig. 2.5 Treacher Collins' clinical presentation (from three different patient angles) with malar hypoplasia, mandibular hypoplasia, absence of zygomatic arch, microtia, atresia of external ear canal, downward slanting palpebral fissures, coloboma of the lower lid, and hypoplasia of lower lid lashes.

syndrome results in abnormal bilateral first and second brachial arch development due to mutations in the gene *TCOF1*.

Treacher Collins syndrome is clinically heterogeneous, but the following common clinical features distinguish patients with this diagnosis: bilateral zygomatic and malar hypoplasia and mandibular hypoplasia, including microgenia, microtia, atresia of external ear canal, downward slanting palpebral fissures, coloboma of the lower lid, hypoplasia of lower lid lashes, cleft palate, conductive deafness, and choanal atresia (► Fig. 2.5). According to Tessier's well-known and cited classification of clefts, Treacher Collins syndrome consists of cleft palate between the 6 through 8 positions. Cleft number 6 refers to oral-ocular clefts, which is seen in incomplete forms of Treacher Collins syndrome; cleft numbers 7 and 8 refer to lateral facial clefts, with number 7 being the most common of all craniofacial clefts. Airway management is a priority in newborns with Treacher Collins syndrome because of the narrow pharyngeal diameter and mandibular shortening, often necessitating intubation and tracheotomy.

In clinically diagnosing Treacher Collins syndrome, it is important to consider acrofacial dysostoses in the differential diagnosis. The absence of limb anomalies differentiates Treacher Collins syndrome from Nager's and Miller's syndromes. Although oculoauriculovertebral (OAV) syndrome has overlapping facial deformities with Treacher Collins syndrome, patients with OAV syndrome are distinguished by hemifacial microsomia, with or without epibulbar dermoids, bilateral anterior accessory auricular appendages, and vertebral abnormalities. These should all be considered carefully in the differential diagnosis, but recognizing that Treacher Collins syndrome is bilateral.

2.3 Nonoperative Management

Nonoperative management is not recommended for these five craniosynostosis-related syndromes: Apert's, Crouzon's, Muenke's, Pfeiffer's, and Saethre-Chotzen, or for Treacher Collins syndrome.

2.4 Operative Treatment

2.4.1 Craniosynostosis-Associated Syndromes

The treatment of five eponymous craniosynostosis-related syndromes, that is, Apert's, Crouzon's, Muenke's, Pfeiffer's, and Saethre-Chotzen syndromes, is surgical management and comprehensive, corroborative, clinical care by a multidisciplinary team, including a plastic surgeon, neurosurgeon, otolaryngologist, ophthalmologist, orthodontist, dentist, pediatrician, clinical geneticist, speech therapist, audiologist, and social worker.

The two main surgical options for craniosynostosis repair are open cranial vault remodeling and minimally invasive surgery. Minimally invasive surgeries, such as strip craniectomy and spring-mediated cranioplasty, are better suited for single-suture synostosis (such as sagittal synostosis) in patients younger than 5 months. Currently, standard of care treatment for syndromic craniosynostosis patients is as follows, in sequence: (1) an initial FOA and cranial vault remodeling, (2) a midface advancement procedure with or without distraction (Le Fort III or monobloc), and (3) secondary orthognathic surgery to correct any dentofacial deformities, that is, Le Fort I with or without mandibular osteotomies. Conventionally, surgical intervention is divided into an early and late timing period in a child's life. Early procedures, usually in the first 4 to 12 months of life, include suture release, cranial vault decompression with or without posterior vault remodeling, and upper orbital advancement and reshaping. Late procedures occurring between 4 and 12 years of age revolve around reconstructing midface retrusion and deformities, whereas in late procedures occurring between 14 to 18 years of life, jaw surgery is the pre-eminent focus.

For early procedures, FOA should be performed during infancy (before 12 months), since it is the period of most rapid cerebrocranial growth, and it corrects fronto-supraorbital

retrusion, protects the globes, reduces compensatory turric-phaly, and provides additional cranial space for the developing brain. Very early correction at less than 6 months of age boasts the advantage of an easily manipulated, malleable fronto-orbital bone. However, later correction between 6 and 12 months is preferable in the authors' opinion, since the cranial bone is more substantial, facilitating rigid fixation, and the vast majority of the abnormal cranial deformity has occurred, thereby lessening the relapse potential following remodeling. Age at initial surgery, length of operation, and estimated blood loss are all cited as not predicative of higher reoperation rates. However, according to Wong et al, although the age at initial FOA did not influence reoperative rates for craniosynostosis, phenotypic syndromic diagnosis was the determinant for outcome, as defined by need for secondary FOA, forehead plasty, or both. Specifically, patients with Apert's syndrome had the highest incidence of reoperation for frontal retrusion or forehead contour, as opposed to patients with Crouzon' and Saethre-Chotzen syndromes, who were most likely to express a minor phenotype and did not require fronto-orbital correction.

However, recently, there has been rising interest in minimally invasive approaches to syndromic craniosynostosis, with techniques including endoscopic suture release, spring-mediated cranioplasty, and distraction osteogenesis. Advocates for minimally invasive surgery cite lower rates of morbidity and mortality as compared with open approaches. In light of this, open repair of craniosynostosis has been increasingly reexamined, with particular attention being paid to the degree of operative blood loss, transfusion requirements, and hospital length of stay, as compared with minimally invasive techniques. However, a recent publication on evidence-based approach to craniosynostosis reported that FOA and cranial vault remodeling are the mainstays of treatment of craniosynostosis. This report by Chim et al in 2011 is corroborated by a very recent large-scale national survey on current practice patterns for craniosynostosis, which was distributed, given the great variability in perioperative management of craniosynostosis. In this survey, completed by 53 craniofacial surgeons, surgeons with greater than 10 years of experience were significantly more likely to perform open repair for craniosynostosis at the extremes of age (<4 months and 8–12 months) and reported significantly shorter operative times compared with their less-experienced peers.

In addition, a retrospective study comparing extended-strip craniectomy and cranial vault remodeling demonstrated that cranial vault remodeling had a significantly higher improvement in the cephalic index. Even in the extended-strip craniectomy cohort that had the procedure in the first 4 months of life, cranial vault remodeling still demonstrated improved results. Although there is a paucity of literature on prospective long-term studies comparing the results of extended-strip craniectomy with molding helmet therapy compared with those of cranial vault remodeling, cranial vault remodeling remains the gold standard and is the authors' preferred technique based on experience and current evidence in the literature.

Regarding technique tips, for surgical access, a zigzag bicoronal incision is best, since it prevents hair parting along a straight line, and the scar tends to spread less due to redistribution of forces. Ideally, the incision should begin slightly anterior and superior to the helix of the ear, and prudent caution should

be taken to not extend the incision at the expense of injury to the temporal branch of the facial nerve. Dissection is done in the subperiosteal plane until the area of the temporalis muscle is reached. Of note, the temporalis muscle is not raised with the coronal flap in order to allow it to be separately dissected and repositioned after FOA. A rim of periosteum surrounding the temporal muscle is left with the muscle during dissection of the coronal flap. The dissection plane continues to be subperiosteal anteriorly (medial to the temporalis muscle), but laterally (over the temporalis muscle), it should be deep to the deep temporal fascia, staying deep to the frontal branch of the facial nerve. As the orbital rim is approached, and anterior to the temporalis muscle, the entire plane becomes subperiosteal again. The periorbita is carefully preserved and dissected away from the bony supraorbital ridge. Small burr holes are utilized for the initial craniotomy, and then, ultimately, using periosteal elevators, the calvarium is elevated off of the dura. The frontal bones are as a bifrontal craniotomy. An approximately 2 cm of supraorbital ridge is left for the fronto-orbital bar, also known as a bandeau. The fronto-orbital bar is removed separately, laterally starting from the frontozygomatic (FZ) suture cephalad on the sphenoid through the lateral portion of the orbital roof, staying anterior to the temporal bone. Medially, the frontonasal suture is separated. The superior orbital rim is separated from the anterior cranial fossa by using an intracranial approach in concert with the neurosurgeon and joining with the lateral orbital and frontonasal cuts. The frontal bones are scored and then molded, for instance, by using starburst craniotomies, and shaped to round out the forehead. The fronto-orbital bar is similarly rounded and is subsequently advanced anteriorly and fixed by rigid fixation. The frontal bones are then secured to the fronto-orbital bar inferiorly and to the parietal cranium posteriorly. Barrel stave osteotomies are used to harvest parietal bone as bone grafts to bridge the gaps after advancement (► Fig. 2.6). Rigid fixation is best achieved with absorbable plates, which are composed of a combination of polyglycolic acid and polylactic acid that absorb by hydrolysis in 1 to 3 years.

Subsequently, distraction of the hypoplastic midface should be performed between 4 and 8 years of age. The decision for when this second phase in the surgical timeline occurs is predicated on the patient's degree of comorbid airway obstruction, OSA, ocular or dental issues, and overall health status.

Le Fort III or the monobloc procedure is done to correct midface retrusion (► Fig. 2.7). A Le Fort III advancement encompasses a complete craniofacial dysjunction and allows movement of the midface only, whereas a monobloc includes the anterior cranium, so that the forehead and midface are moved in the same operation. Although advantageous for its simultaneous correction of the forehead and midface deformity, the monobloc procedure is associated with greater blood loss, risk of cerebrospinal fluid (CSF) leak and higher infection rate, due to the nature of direct communication between the cranial and nasal cavities.

Briefly, in a Le Fort III advancement, the lateral osteotomies are as follows: (1) the FZ suture carrying inferiorly to separate the greater wing of the sphenoid and the zygoma (lateral orbital wall) to the inferior orbital fissure, and (2) the zygomatic arch is osteotomized to fully mobilize the zygoma. Inferior and posterior to the FZ suture, the osteotomies include the pterygomaxillary junction. Medially, the osteotomies include the frontonasal

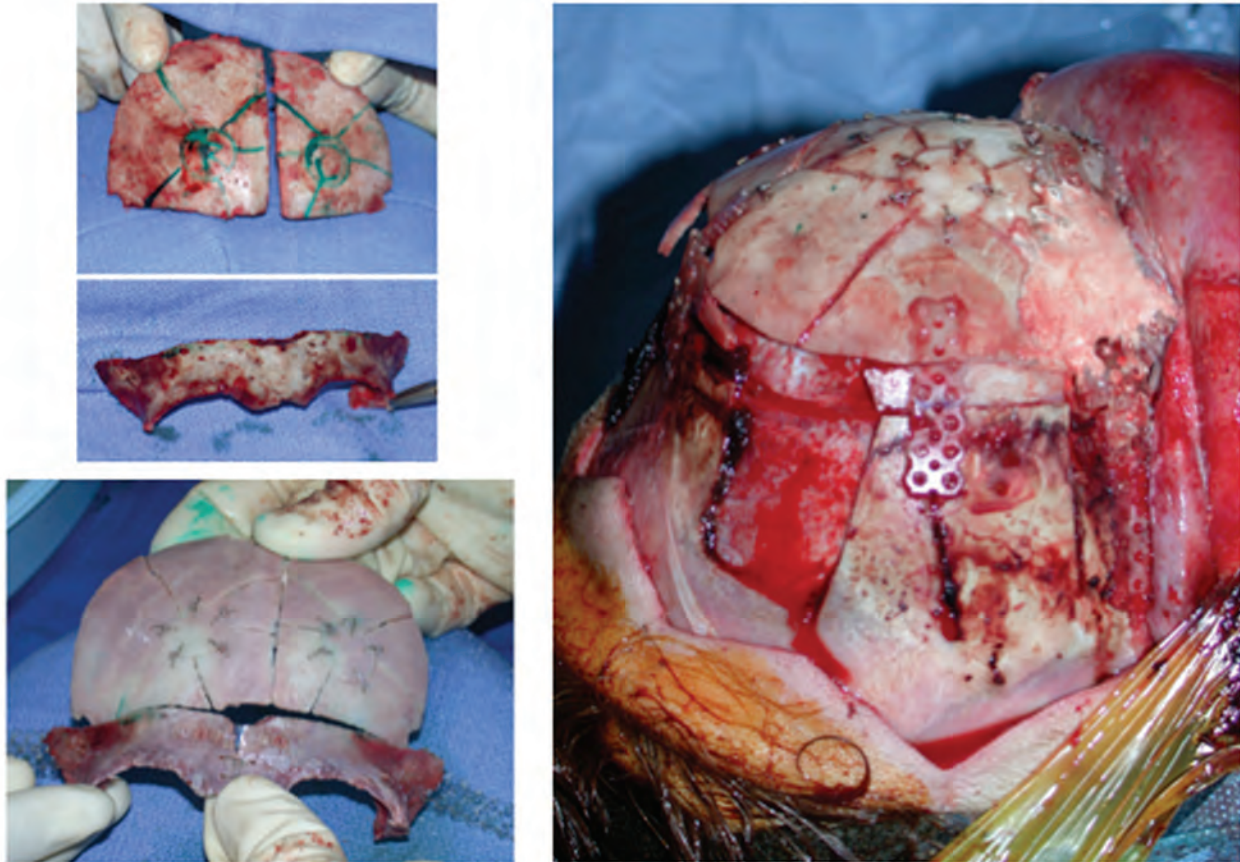


Fig. 2.6 Fronto-orbital advancement (FOA) intraoperative photographs.

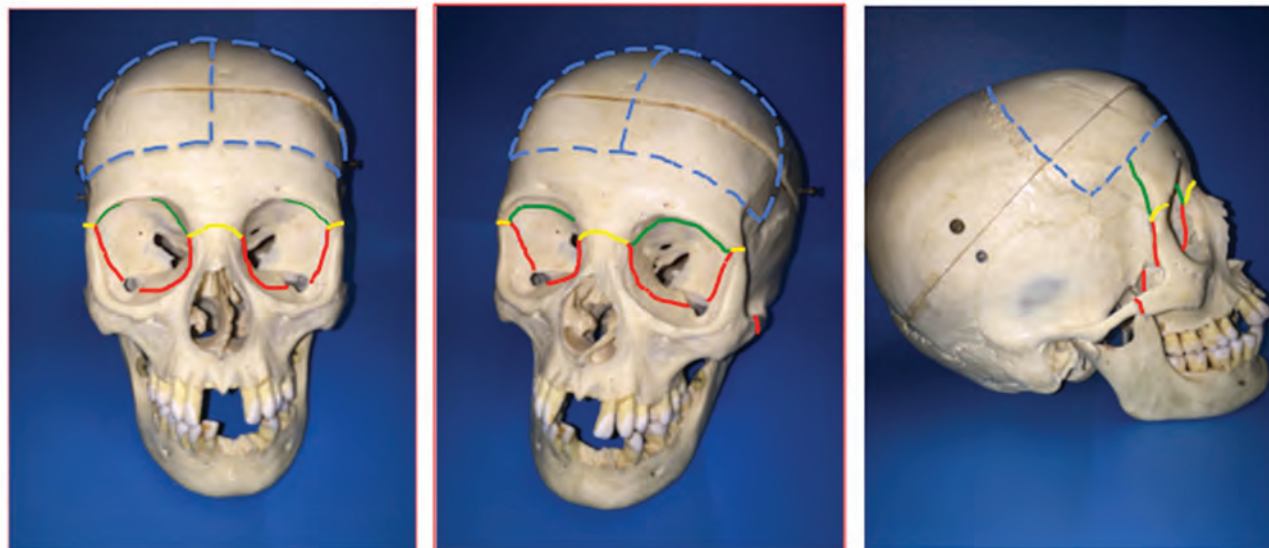


Fig. 2.7 Osteotomies for Le Fort III (yellow and red) and monobloc advancement (green and red). In monobloc advancement, the frontal bones are also removed in order to provide access to the orbital roof. The dashed blue line denotes the frontal craniotomy for access to the orbital roof.

junction, which is carried down posterior to the lacrimal fossa in order to separate the ethmoid and lacrimal bones, thereby facilitating greenstick fracture and en bloc movement of the ethmoid and lacrimal bones contiguous with the midface (medial orbital wall and orbital floor). Prudent care is taken to not disrupt the medial canthus and lacrimal apparatus. Subsequently, the ethmoid and vomer are osteotomized and separated from the cranial base. Then, the pterygomaxillary junction is osteotomized and greenstick fractured, which allows for the final step of the midface being down fractured and mobilized.

In a monobloc advancement, the frontal bones are mobilized with the midface. In contrast to the Le Fort III advancement, the fronto-orbital bar is included with the midface for mobilization. First, the approach begins with a bifrontal craniotomy to allow access to the orbital roof and crista galli. Prudent care must be taken to remain anterior to the crista galli in order to minimize risk of damage to the olfactory nerves or incurring a CSF leak. The orbital roof is separated from the cranial base via an intracranial approach. The osteotomy is carried laterally to include a portion of the lateral frontal bone posterior to the lateral orbital rim. The FZ and frontonasal sutures are left intact. Overall, the osteotomies for the lateral and medial orbital walls and the orbital floor, the pterygomaxillary junction, and the ethmoid/vomer separation from the cranial base are similar to the Le Fort III procedure.

Finally, orthognathic surgery is the last phase of reconstruction, with a Le Fort I osteotomy or bimaxillary surgery being utilized to achieve normal occlusion.

2.4.2 Craniosynostosis-Associated Syndromes: Syndrome-Specific Considerations

All of the aforementioned craniosynostosis syndromes have abnormal, premature fusion of one or more cranial sutures. However, Apert's and Pfeiffer's syndromes have prominent midface retrusion, whereas the midface position is less severely affected in other conditions such as Muenke's and Saethre-Chotzen syndromes. Surgical and nonsurgical management will have to be tailored based not only on the genetic and clinical diagnoses but also on the severity of symptoms, presence of midface hypoplasia, and functional compromise experienced by the patient. For these craniosynostosis syndromes, it is best to categorize specific management considerations by the presence or absence of midface hypoplasia.

For both Apert's and Pfeiffer's syndromes, the treatment is surgical management and comprehensive, corroborative care by a multidisciplinary team, including a plastic surgeon, neurosurgeon, otolaryngologist, ophthalmologist, orthodontist, dentist, pediatrician, clinical geneticist, speech therapist, audiologist, and social worker.

For Apert's syndrome, FOA and cranial vault remodeling in infancy at about 9 months of age (age range, 9–12 months) are the current standard for treatment. Bony fixation is best achieved with biodegradable plates and screws, as opposed to titanium plates and screws. Clinically, 95 to 100% of patients with Apert's syndrome who had undergone FOA and cranial vault remodeling in infancy will need reoperation, and forehead

recession after initial reconstruction occurs. Allam et al present one of the longest available follow-up studies for surgical correction of patients with Apert's syndrome, with a mean follow-up of 22.5 years following FOA, and provide invaluable insight into psychosocial integration of patients with Apert's syndrome into society.

Both forehead recession and midface hypoplasia contribute to the functional and aesthetic considerations in the treatment of Apert's syndrome. In the presence of both forehead and midface deformities, monobloc advancement and asymmetric facial bipartition is the preferred operation. If only a midface deformity is present, a subcranial Le Fort III operation is the preferred procedure. Given the complex facial and cranial deformities defining Apert's syndrome, parents may advocate for earlier midface advancement at the age of 4 or 5 years. However, it is advantageous to consider waiting until the age of 6 or 7 years, when mixed dentition has been achieved and most children have reached cranial vault and orbit sizes similar to adult size. However, it must be fully recognized by both the treating team and patients' parents that surgical midface advancement does not lead to normal growth thereafter in children with Apert's syndrome. Thus, additional surgical procedures needed can include Le Fort I osteotomy, nasal dorsum augmentation, myringotomy, and surgical contouring over the superficial temporal fat pads due to hypoplasia. It is important to note that surgical interventions for the treatment of Apert's syndrome are associated with relatively low complication rates; mean infection rate is 2.3% following cranial vault procedures and 6.5% following midface advancements.

Regarding management of the pathognomonic pan-syndactyly involving both the hands and feet, treatment can be separated into two phases: early (syndactyly releases) and late (functional osteotomies). As described recently by Fearon, improved surgical treatment of Apert's hands and feet now involves the following practices: (1) the release of all 10 fingers and toes in only two operations, (2) elimination of routine digital amputations, (3) adoption of the straight-line release and abandonment of the zigzag incision, (4) leaving of small areas of exposed bone without vascularized tissue coverage, and (5) performance of mid-phalangeal osteotomies in older children to improve hand function.

For Pfeiffer's syndrome specifically, elevated intracranial pressure is more common with multisuture craniosynostosis, and it is an important consideration in management, especially with type II featuring cloverleaf skull (kleeblattschadel). Coordination between the neurosurgeon and plastic surgeon is vital to complete cranial vault expansion in the first year of life in order to allow the possibility for normal brain development.

As with all syndromic craniosynostosis, Pfeiffer's syndrome should be treated in the first 9 to 12 months of age with an anterior cranial vault expansion by FOA and cranial vault remodeling. Ultimately, the objectives of early cranial vault surgery and FOA in this patient population are to provide adequate repositioning of the forehead and cranium, to facilitate sufficient volume for brain development, and to offer ocular protection. Subsequently, midface hypoplasia is usually corrected in late childhood or early adolescence by Le Fort III osteotomy or monobloc advancement. Finally, orthognathic surgery is the last phase of reconstruction with a Le Fort I osteotomy or bimaxillary surgery to achieve normal occlusion.

For continuity of care in the management of both Apert's and Pfeiffer's syndromes, multidisciplinary team follow-up should be scheduled for the patient at 1 month postoperatively and subsequently once every 3 months after each major surgical procedure. This is the minimum requirement for follow-up with the team, and most patients require more frequent visits in the first 6 months postoperatively. Thereafter, follow-up appointments should be scheduled annually until late adolescence. Treatment goals should be focused on preventing avoidable developmental delays secondary to increased intracranial pressure and OSA, minimizing the number of operative interventions, and achieving normalized skeletal appearance by skeletal maturity. Patients and patients' parents should be asked about any respiratory difficulties, ocular complaints, hearing issues, signs, and symptoms suggestive of increased intracranial pressure as well as psychosocial and educational development. Skull circumference measurements, facial feature assessments, and radiographs should be obtained at regular intervals. Patients' parents should be counseled appropriately on the need for ongoing follow-up and the prospect of multiple reoperations to account for the multifactorial nature of these craniofacial syndromes.

Furthermore, for Crouzon's, Muenke's, and Saethre-Chotzen syndromes, the treatment is surgical management and comprehensive, corroborative care by a multidisciplinary team, including a plastic surgeon, neurosurgeon, otolaryngologist, ophthalmologist, orthodontist, dentist, pediatrician, clinical geneticist, speech therapist, audiologist, and social worker.

As with all syndromic craniosynostosis, Crouzon's, Muenke's, and Saethre-Chotzen syndromes should all be treated in the first 9 to 12 months of age with an anterior cranial vault expansion by FOA and cranial vault remodeling. Ultimately, the objectives of early cranial vault surgery and FOA in this patient population are to provide adequate repositioning of the forehead and cranium, to facilitate sufficient volume for brain development, and to offer ocular protection. Although much less common with these three syndromes, midface hypoplasia, if present, is usually corrected in late childhood or early adolescence by Le Fort III osteotomy or monobloc advancement. Finally, orthognathic surgery is the last phase of reconstruction with a Le Fort I osteotomy or bimaxillary surgery to achieve normal occlusion.

Notably, patients with Muenke's syndrome have the lowest incidence of cleft palate among the craniosynostosis syndromes. However, there is a very high incidence of high-arched palate in Muenke's syndrome, which merits close follow-up and treatment. Because of this high-arched palate and likely interference with the tensor veli palatini muscle, chronic otitis media occurs frequently in this patient population. Hence, patients with Muenke's syndrome can develop recurrent chronic otitis media with effusion and hearing loss, and development of these sequelae should be monitored as part of the treatment plan.

Finally, Crouzon's, Muenke's, and Saethre-Chotzen syndromes have extensive psychosocial and aesthetic considerations, and management necessitates a multidisciplinary team approach in close communication with patients' parents. Routine postoperative follow-up visits should be scheduled at 1 month and subsequently once every 3 months after a major procedure for craniosynostosis repair. Then, the patient should follow up with the craniofacial team every year until late adolescence for ongoing evaluation.

2.4.3 Cleft-Associated Syndrome

The treatment of Treacher Collins syndrome is surgical management and requires collaboration among a multidisciplinary team, including a plastic surgeon, neurosurgeon, otolaryngologist, ophthalmologist, orthodontist, anesthesiologist, dentist, pediatrician, clinical geneticist, speech therapist, audiologist, and social worker. The key to the surgical management of these patients is intervention of a cross-disciplinary team and correct surgical timing for the various stages of reconstruction, with consideration for both physical and mental development.

Although airway management is the initial priority, often with tracheostomy, the subsequent reconstructive surgeries proceed in a staged, timely fashion. First, within 9 to 12 months after birth, surgeries alleviating feeding difficulties are performed, namely glossopepy and cleft palate repair. Of note, the repair technique utilized for these cleft palates should be the same as those used for routine nonsyndromic cleft cases. Starting from the age of 3 years to adolescence, patients with Treacher Collins syndrome who have cleft palates should work closely with a speech therapist to maximize speech function and outcomes. Reconstruction of the orbitozygomatic region should be performed at approximately 8 to 10 years of age with autologous bone grafts. For ocular reconstruction, lateral canthoplasty can be secured to the reconstructed malar complex, and lower eyelid colobomas can be addressed with a pedicled upper to lower eyelid musculocutaneous flap. Third, external ear construction for microtia is performed at approximately 8 years of age or older. The four-staged surgical technique for the ear, described by Brent, which begins with the construction of an autologous rib cartilage framework, is the authors' preference for treatment. Other ear construction methods that are commonly used include variations of autologous costal cartilage grafts, namely the Nagata's two-stage technique, the Walton and Beahm's technique, the Tonzor's technique, tissue expansion, implants, osseointegration, and prostheses. Finally, orthognathic surgery should be completed between the ages of 16 and 18 years following achievement of skeletal maturity. Of note for the treatment of malocclusion, if the temporomandibular joint is absent, a costochondral graft should be placed at approximately 6 to 10 years of age and then subsequently addressed with further orthognathic surgery in late adolescence.

2.5 Complications

Possible surgical complications following cranial vault remodeling include the following:

- Hemorrhage.
- Infections.
- Injury to the dura.
- Bony contour irregularity.
- Bony defect.

Possible surgical complications following Le Fort III and monobloc frontofacial advancement are similar. However, given its added intracranial approach and the inherent communication between cranial and nasal cavities, the monobloc procedure carries added risk of intracranial injury, CSF leak, and infections overall. Complications for Le Fort III include the following:

- Globe injury.
- Optic nerve injury.
- Ocular exposure.
- Stripping of the medial canthal tendon.
- Enophthalmos.
- Temporal fat atrophy.
- Hemorrhage.
- Infections.
- Inadvertent extubation.
- Injury to developing or permanent dentition.
- Hardware failure (if distraction is utilized).
- Hardware infections (if distraction is utilized).

In addition to these, the monobloc procedure has added risk of the following:

- CSF leak: common occurrence (2–20% of cases) but usually self-resolving.
- Frontal bone flap loss: 2%; however, rates vary widely in the literature.
- Infections: Due chiefly to the inherent communication between intracranial and nasal cavity, with lower rates found for distraction osteogenesis.
- Mortality: Uncommon (0–4.5%, with most recent series quoting rates under 1%).
- Mucocoele: Long-term risk.

Possible surgical complications following microtia repair include:

- Infection, in most severe case can lead to total loss of cartilage graft.
- Skin envelope viability and healing, may need salvage with temporoparietal fascial flap and full thickness skin graft.
- Hematoma.
- Seroma.

2.6 Conclusion

Craniofacial syndromes can be divided into two major categories: craniosynostosis- and cleft-associated syndromes. The diagnosis and management of syndromic craniofacial anomalies are complex due to the overlapping and variable phenotypic presentations. Common forms of syndromic craniosynostosis include Apert's, Crouzon's, Muenke's, Pfeiffer's, and Saethre–Chotzen syndromes, whereas Treacher Collins syndrome is the most common cleft-associated mandibulofacial dysostosis condition. Management of patients with craniofacial syndromes includes accurate diagnosis, correctly timed surgical operations in a staged fashion, close involvement of the patients' families, as well as psychosocial and aesthetic considerations for the pediatric patients. A multidisciplinary team approach is vital to a successful outcome, and the involvement of a pediatric plastic surgeon in the surgical management of these complex craniofacial syndromes is central.

2.7 Key Points

- Common forms of syndromic craniosynostosis include Apert's, Crouzon's, Muenke's, Pfeiffer's, and Saethre–Chotzen syndromes, whereas Treacher Collins syndrome is the most

common cleft-associated mandibulofacial dysostosis condition.

- All of the craniosynostosis syndromes discussed (Apert's, Crouzon's, Muenke's, Pfeiffer's, and Saethre–Chotzen) have abnormal, premature fusion of one or more cranial sutures. However, Apert's and Pfeiffer's syndromes have prominent midface retrusion corrected by Le Fort III or monobloc procedure, whereas the midface position is less severely affected in other conditions such as Muenke's and Saethre–Chotzen syndromes. Surgical management will have to be tailored based not only on the genetic and clinical diagnosis but also on the severity of symptoms, presence of midface hypoplasia, and functional compromise experienced by the patient.
- The two main surgical procedures for craniosynostosis repair are FOA with open cranial vault remodeling and strip craniectomy, which can be performed in a minimally invasive endoscopic-assisted fashion. The goals of surgical treatment and overall management of patients with Treacher Collins syndrome include palate repair, speech therapy, construction of ears, reconstruction of the zygomatic prominence, and correction of midface retrusion to improve the airway and respiratory status as well as to improve occlusion.
- Management of patients with craniofacial syndromes includes accurate diagnosis, correctly timed surgical operations in a staged fashion, close involvement of the patients' families, and psychosocial and aesthetic considerations for the pediatric patients. A multidisciplinary team approach is vital to a successful outcome.

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3 Deformational Plagiocephaly, Brachycephaly, and Scaphocephaly

Gary F. Rogers and Benjamin C. Wood

Summary

Deformational plagiocephaly is a common condition that affects infants. Patients have a head preference when supine, usually because of torticollis. As a result, the cranium becomes misshaped over the first few months of life. Treatment includes positioning strategies or cranial molding device.

Keywords: deformation, torticollis, helmet, plagiocephaly, brachycephaly, scaphocephaly

3.1 Introduction

In 1992, the American Academy of Pediatrics initiated the “Back to Sleep Campaign” in an effort to reduce the incidence of sudden infant death syndrome. This policy advocated a paradigm shift of infant sleep positioning in Western countries from prone to supine. The recommendation was widely implemented and resulted in an estimated 40% reduction in the incidence of sudden infant death syndrome in the United States. One of the unforeseen consequences of the campaign was a rise in deformational cranial flattening and asymmetry. Such alterations of cranial shape were less commonly seen in prone-slept infants and were very common in certain Eastern countries that historically placed their sleeping infants supine. However, precipitous rise in deformational changes in the early 1990s sparked much concern from clinicians and parents alike. Several large craniofacial centers even misdiagnosed this condition as lambdoidal craniosynostosis and began undertaking widespread operative correction. The confusion eventually cleared; however, the diagnosis and treatment of plagiocephaly remain a topic of much debate. According to some estimates, the prevalence is as high as 20% in healthy infants.

3.2 Diagnosis and Terminology

The literature is full of incorrect terminology related to deformational flattening. Many authors and clinicians refer to every form of cranial flattening as “plagiocephaly.” This generally applied term is nonspecific. Deformational cranial flattening can be asymmetric, symmetric, or a combination. Plagiocephaly is derived from the Greek words *plagios*, meaning oblique or slanted, and *kephalē*, meaning head (► Fig. 3.1). This condition occurs primarily in infants who consistently favor turning their head to one side, that is, those with congenital muscular torticollis (CMT). The resultant cranial shape has been compared to a “parallelogram,” but the frontal asymmetry is less severe than what is observed in the occiput, and the shape is more trapezoidal. Asymmetric growth of the head often is accompanied by facial asymmetry—anterior (sagittal) shift of the ipsilateral forehead, ear, and cheek. Plagiocephaly is the most common cause of clinically evident facial asymmetry in infants and should be included in the differential diagnosis for any child with such findings.

Brachycephaly (Greek word *brachy* = short) denotes symmetrical occipital flattening and compensatory parietal widening. Infants with this condition have little or no rounding on the back of the head and appear to have a disproportionately wide or “big” head when viewed from the front (► Fig. 3.2). The posterior vertex may appear taller than the front (turricephaly), giving a sloped appearance to the head in profile. The cranial width-to-length ratio, termed the cranial or cephalic index (CI), is generally higher than normal (>0.85). Most children with this condition also have some element of concurrent asymmetry, or plagiocephaly. The combination effect, termed “asymmetric brachycephaly,” is the most common type of deformational shape.

Deformational scaphocephaly (“boatlike head”) is an uncommon variant of plagiocephaly (► Fig. 3.3). It is more commonly



Fig. 3.1 Right posterior deformational plagiocephaly.



Fig. 3.2 Deformational brachycephaly.



Fig. 3.3 Deformational scaphocephaly.

seen in infants who have extreme head rotation to one side or in premature infants that are positioned side to side in the intensive care units. Flattening develops on the side(s) of the head, and compensatory expansion occurs in the anterior and posterior cranium. These infants tend to develop a long, slender head, colloquially referred to by some as a “toaster head.” There is often relatively pronounced facial asymmetry. This presentation can be confused with scaphocephaly caused by sagittal craniosynostosis, or premature fusion of the sagittal suture. In contrast to deformational scaphocephaly, sagittal craniosynostosis typically results in frontal bossing, bilateral occipital/parietal narrowing *posterior* to the anterior fontanelle, and decreased vertical height

of the posterior cranium. Facial asymmetry is rare in sagittal synostosis. Most infants with this type of craniosynostosis have a head circumference in excess of the 90th percentile.

3.2.1 Radiographic Imaging

In general, any child with an abnormal head shape should be referred to a specialist *before* ordering radiographic studies. The overwhelming majority of infants with cranial asymmetry will have deformation and not synostosis, and a specialist can usually distinguish these processes from craniosynostosis by history and physical examination alone. This approach avoids

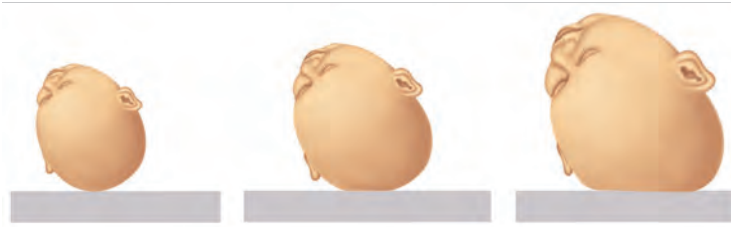


Fig. 3.4 Mechanism of flattening; progressive cranial expansion against a flat surface.

exposing infants to unnecessary radiation and possible anesthetic exposure. If imaging is deemed necessary, the specialist is best suited to determine what is needed, based on the diagnostic need. The accuracy of plain radiography to diagnose suture fusion is questionable. Computed tomography (CT) is costly, often requires sedation, and involves low-dose ionizing radiation. The impact of such exposure is controversial; however, theoretical studies suggest that even a low-dose CT in an infant can increase the risk of lethal brain cancer.

3.3 Etiology

Deformational changes are thought to arise predominantly in the postnatal period in response to external forces between the sleeping surface or bed and the growing infant cranium. The cranium grows passively in response to minor internal pressure exerted by the rapidly growing infant brain. This process is fastest in early infancy and tapers dramatically even after the first year of life. When an infant is placed on a resting surface, there is a contact force generated between the head and the surface (Newton's third law). If the point of contact is not changed, the counterforce will restrict cranial growth in the area of contact, and over time, any volume increases will be displaced to areas where there is no resistance (► Fig. 3.4). This is analogous to how a pumpkin flattens as it grows in a field—it cannot expand into the ground and must grow parallel to it. Most parents of affected children begin to notice head flattening in their infants at an average of 6 to 8 weeks of age, because it takes this long for cranial flattening to manifest. The compensatory and redirected growth will result in progressive flattening. Fortunately, most infants have sufficient neck range of motion and strength to alter (or allow the parent to alter) their head position and the area of contact with the bed. Those who lack this capacity are at greatest risk for developing flattening.

The reasons why some infants are able to change their head position (and avoid flattening) while others are not is worth discussion. Numerous risk factors have been linked to the development of deformational flattening, including supine sleep position, multiple births, developmental delay, small maternal pelvis, breech position, oligohydramnios, male gender, gestational diabetes, nulliparity of mother, high birth weight, large neonatal head size, vaginal delivery, prolonged length of postdelivery hospital stay (> 4 days), and prolonged duration of stage II labor. Although these variables seem unrelated, they share a common pathogenic link to the development of deformational cranial flattening: each directly or indirectly impairs infant head mobility early in life. For example, most of the risk factors described earlier are also associated with the development of CMT, the most common risk factor for deformational

flattening. Congenital muscular torticollis is not always easy to detect in a newborn, but the presence of a “preferred” head position early in life is highly suggestive. Infants with CMT have restricted motion in one direction and excessive head rotation in the other. This is an imbalance of the sternocleidomastoid muscles from in utero positioning—one contracted and the other stretched and atrophied. The characteristic head position is tilted to one side (toward the side of the tight muscle) and rotated to the other (toward weaker muscle). In most infants, increased motor development allows gradual resolution of CMT. The timing of improvement can vary based on the severity of the contracture or imbalance and the neuromuscular development of the infants. Once the infant can move his or her head, usually at 4 months of age, further flattening is unlikely.

The two other major etiologic groupings, prematurity and developmental delay, can also increase the risk of developing cranial flattening because they delay the development of independent head mobility. In infants who have both CMT and one of these other variables (e.g., twins), the risk of developing deformational flattening is markedly increased.

3.4 Assessment

Direct or indirect anthropometry offers some objective standard on which to predicate treatment. Direct measurements can be easily obtained using an anthropometric caliper (► Fig. 3.5). Deformational brachycephaly (DB) is almost always quantified by using CI, which is the maximum width of the head divided by the anteroposterior length (► Fig. 3.6). Deformational plagiocephaly (DP) is quantified by the degree of asymmetry, using either absolute measurements (the difference between two oblique head measurements; ► Fig. 3.7) or cranial ratios (the ratio of one oblique to the other, or the absolute difference divided by head length). The use of ratios to assess asymmetry is inherently flawed, since any degree of fixed asymmetry will appear to “improve” as the head grows in total size. Surface laser scans are also now widely used and are a reasonable proxy for direct anthropometry. This method also provides a three-dimensional visual of the head shape.

3.5 Prevention and Treatment

3.5.1 Infants at Risk: Early Identification and Intervention

The best opportunity to prevent deformational flattening is to identify at-risk infants, and the most important piece of information is whether or not the infant has a head positional preference. This simple observation is the key to identifying most

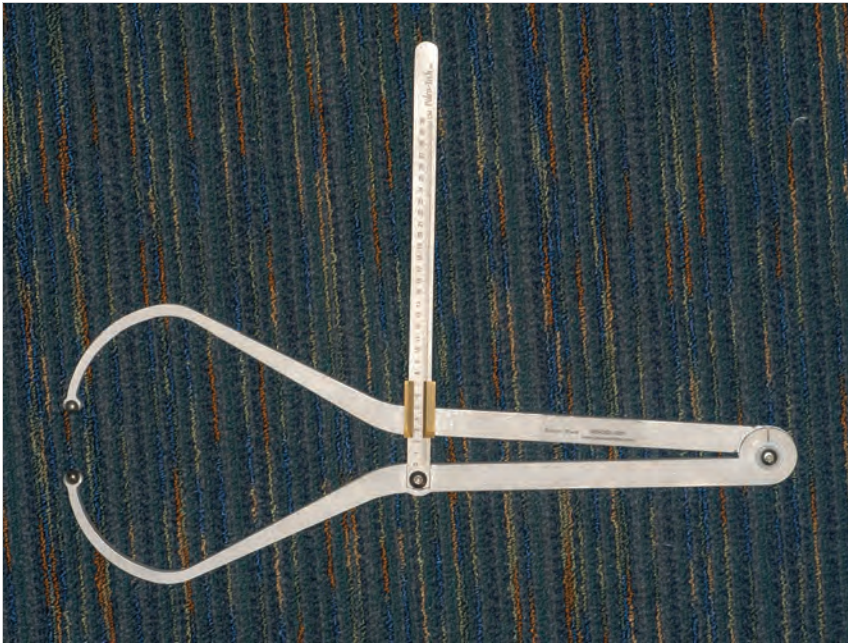


Fig. 3.5 Cranial caliper for direct anthropometry.

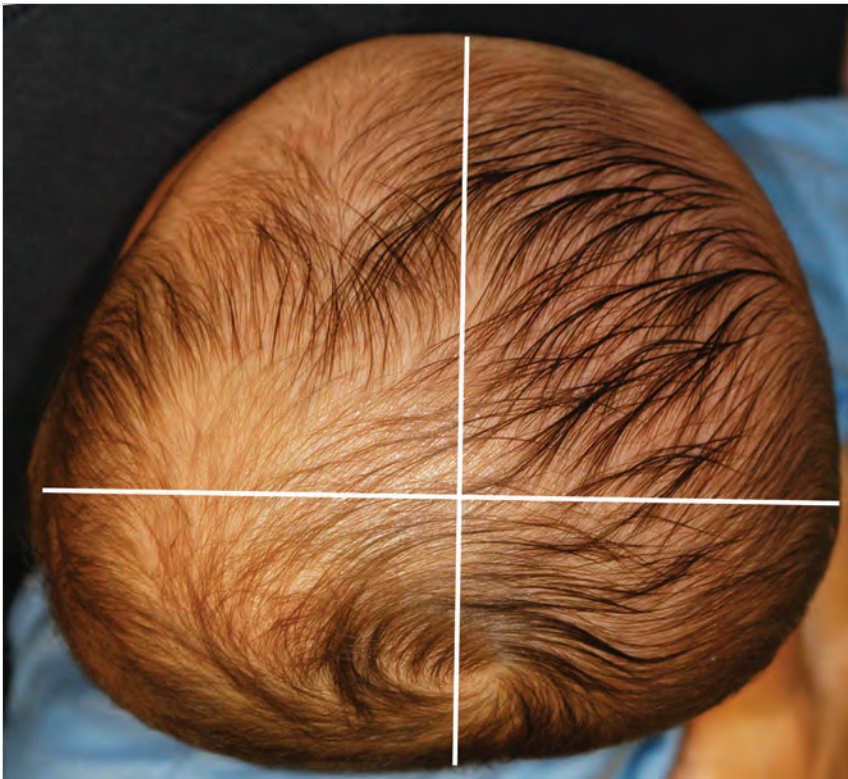


Fig. 3.6 Measurement of cranial index (width/length).

vulnerable infants. Traditional recommendations for infants who appear at risk for DP or DB, or have just started to develop a flat spot, include physical therapy to address the cervical muscular contracture and repositioning. Data to support these recommendations are sparse. Repositioning is possible only in very young and immobile infants (<4 months of age) and, if done consistently, may serve to prevent or limit further deformation. This method is extremely difficult to use in older, mobile infants

and is generally ineffective at correcting established flattening. Sleep surface alterations have shown promise in preventing and/or correcting plagiocephaly in infants younger than 4 months.

3.5.2 Older Infants

By the age of 4 months, most infants begin to develop head control and are mobile enough that they should not develop

further flattening. The vast majority of infants with torticollis will have improved to a point where their head rotation is unrestricted and the concurrent head tilt may be subtle, intermittent, or completely absent. Most infants can easily move off wedges and other such devices, and the only option is either continued observation or a helmet orthotic. Indications for treatment are quite variable, and most clinicians make this decision based on subjective impression of severity or some arbitrary measure of severity (e.g., < 12 mm transcranial difference or cranial index of < 0.90). If left untreated, continued

growth of the cranium tends to mitigate the severity of the flattening since a fixed amount of flattening on a larger head will look relatively less pronounced. However, one need not look far to find an adult or an older child with persistent symmetric or asymmetric occipital flattening will appear less pronounced (► Fig. 3.8). Asymmetry will also appear less obvious as hair grows and as the child grows taller because the posterior contour is less obvious when viewed from the posterior perspective (as in an adult) than from a top view (as one often sees infants and young children).

Although an association between deformational flattening and developmental delay has been reported, it is important to understand that it is the neurocognitive delay that leads to an increased risk of cranial flattening (presumably due to delayed motor development and voluntary head repositioning). There is no evidence that cranial flattening affects cognitive or motor development. There is no convincing evidence that deformational flattening causes impairment of visual development or temporomandibular joint (TMJ) function, malocclusion, or other medical conditions.

3.5.3 Helmet Therapy

Helmet orthosis is a useful, and often the only, way to improve moderate and severe deformational flattening. These devices have an excellent track record of safety and effectiveness and are very well tolerated by infants (► Fig. 3.9). A common misconception is that the helmet is actively “molding” or squeezing the cranium and will cause discomfort or limit brain growth. Helmets do not compress the cranium and act more like a brace to redirect remaining cranial growth toward the flattened areas (► Fig. 3.10). A few reports have concluded that helmets are no more effective than observation alone, but these investigations suffer from methodologic flaws (i.e., using oblique cranial ratios instead of absolute measurements) that render these

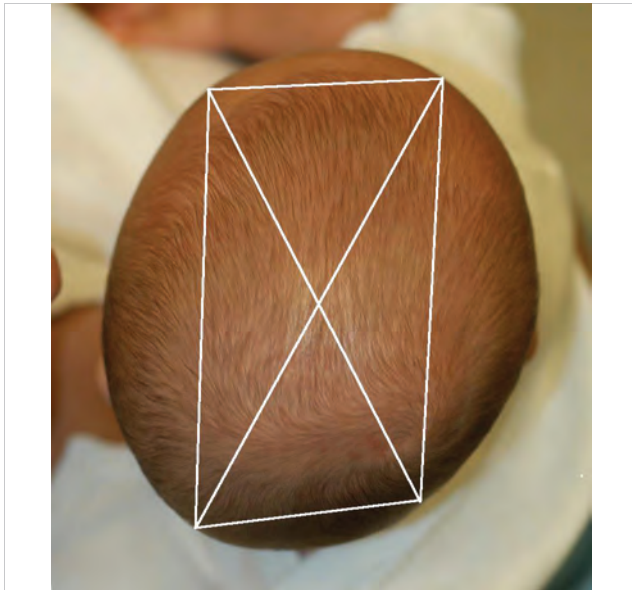


Fig. 3.7 Measurement of transcranial difference (long oblique–short oblique).



Fig. 3.8 A 10-year-old male child with severe persistent right posterior plagiocephaly (Transcranial difference (TCD) = 21 mm; normal 3–5 mm). (a) Vertex view: the asymmetry is pronounced when viewed perpendicular to the plane of the discrepancy. (b) Posterior view: the asymmetry is difficult to detect when viewed in the plane of the asymmetry.



Fig. 3.9 Cranial orthotic.

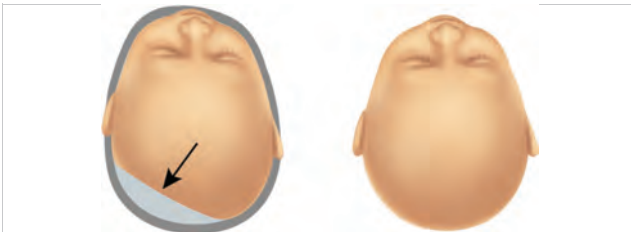


Fig. 3.10 Mechanism of correction by using a cranial orthotic.

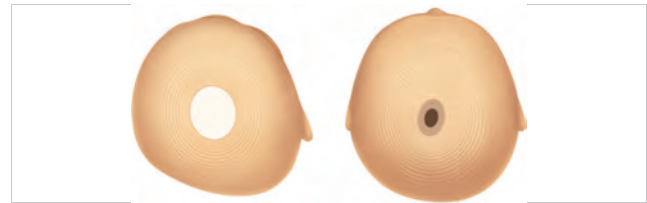


Fig. 3.11 Laser scan of a 6-month-old infant with left posterior plagiocephaly (a) before and (b) after 3 months of helmet therapy.

conclusions unfounded. Furthermore, the effectiveness of any helmet orthosis is dependent on many variables that cannot be easily standardized, such as the age at initiation (better and faster for younger patients), the helmet design (very important), and the duration of and compliance with wear. Since helmets rely on cranial growth for shape improvement, the effectiveness diminishes in older children, and significant improvement is very difficult after 18 months of age. A well-fitted helmet in a younger patient can yield impressive improvements in shape in a relatively short time (► Fig. 3.11).

3.6 Conclusion

Deformational cranial flattening is a benign process that characteristically occurs over the first few months of life as a result of poor infant head mobility. Although many risk factors have been identified, nearly all have some impact on the ability of the newborn to reposition his head early in life. Most affected infants demonstrate a “preferred” head position, which is the earliest evidence of cervical tightness or imbalance. These infants should be considered at risk, and proactive preventative measures should be instituted. Older children who have more pronounced flattening that is conspicuous from a posterior or side view may be considered for helmet therapy. There is little

evidence that untreated cranial flattening has cognitive or medical implications, and cranial flattening present in infancy is mitigated by subsequent cranial enlargement (proportionate improvement) and hair growth.

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4 Craniofacial Microsomia

Patrick A. Gerety, Albert K. Oh, and Jesse A. Taylor

Summary

Management of Mandibular Hypoplasia in Pediatric Unilateral Craniofacial Microsomia attempts to succinctly describe the major principles of the surgical treatment of mandibular asymmetry in patients with unilateral craniofacial or hemifacial microsomia. This chapter also delves into some of the important controversies regarding the surgical management of mandibular hypoplasia, particularly in regards to long-term results.

Keywords: craniofacial or hemifacial microsomia, mandibular hypoplasia/asymmetry, distraction osteogenesis, graft, reconstruction

4.1 Introduction

Hemifacial or craniofacial microsomia (CFM) represents a wide group of malformations primarily involving structures derived from the first and second branchial arches. With reported incidences ranging from 1 in 3,000 to 1 in 20,000, this heterogeneous disorder of craniofacial morphogenesis represents the second most common craniofacial anomaly after cleft lip/palate. Although the etiology of CFM is unclear, two major theories include vascular disruption of the stapedia artery that supplies the first and second pharyngeal arches and abnormal migration of key neural crest cells during embryogenesis. The latter theory provides a more plausible explanation for the bilateral craniofacial features and extracraniofacial findings often seen in expanded-spectrum CFM.

4.2 Diagnosis

With the ongoing advancements in ultrasonography and prenatal magnetic resonance imaging (MRI), CFM can be diagnosed prenatally. However, it is much more common for clinical suspicion to arise postnatally. Abnormalities of the external ear, facial asymmetry, and facial paralysis are most likely to be identified by neonatologists and pediatricians.

4.2.1 Classification System

Although several systems exist for classifying CFM, the OMENS-Plus classification is the most frequently utilized. The OMENS-Plus classification (► Fig. 4.1) succinctly describes and grades the five major craniofacial abnormalities of CFM, specifically involving the (1) orbit, (2) mandible, (3) ear, (4) nerve (facial), and (5) soft tissue deficit. Scores range from 0 (normal) to 3 (most severe). The plus designation applies to extracraniofacial anomalies, with the most common associated abnormalities involving the central nervous, skeletal, and cardiac systems. Patients with more severe CFM (i.e., those with higher OMENS score) have significantly greater risk of having noncraniofacial anomalies, lending further support to the theory of neural crest involvement in the etiopathogenesis of CFM. The clinical

features and their severity will be discussed as follows in the order of the OMENS-Plus classification system.

4.2.2 Orbit

The orbit is affected in a minority (20–30%) of patients with CFM. Orbital manifestations of CFM include a reduction in the dimensions (O1), inferior positioning resulting in vertical orbital dystopia (O2), and, in severe cases, a severely constricted orbit (O3) featuring microphthalmia and hypoplastic lids. Patients with microphthalmia often do not have vision in the affected eye (► Fig. 4.2).

4.2.3 Mandible

Mandibular hypoplasia is the hallmark of CFM and affects 90 to 100% of patients. The mandibular component of CFM has received the most attention in the literature. This is related to surgical innovations (distraction osteogenesis [DO]) and the degree of facial normalization that mandibular surgery can produce. The morphology of the CFM mandible is quite variable (► Fig. 4.3), and there has been a vigorous debate about the best way to classify the patients. Although there are several classification schemes, the Kaban-modified Pruzansky's system is the most commonly used: type I is a small mandible with normal morphology; type IIa is a mandible with a ramus that is abnormal in size and shape but with a normal temporomandibular joint (TMJ); and type IIb is a mandible with an abnormal ramus and TMJ. Type III deformity is defined by an absent ramus, condyle, and TMJ.

In general, the CFM mandible is smaller than the contralateral normal mandible. Much of this size difference is seen in the condyle–ramus portion of the mandible. In mild cases, the effect on the ramus is largely vertical shortening, resulting in decreased posterior facial height and occlusal canting upward on the affected side. In severe cases, specific portions of the mandible, including the condyle, the ramus, the glenoid fossa, and the TMJ, may be absent or unrecognizably distorted. An important observation is that the affected condyle and ramus are shifted medially, contributing to decrease in transverse facial width.

Because moderately and severely affected mandibles are so morphologically affected, classification systems have been found to be unreliable, even among expert raters. Hypoplasia of the maxilla was long thought to be a part of CFM, but vigorous examination has not shown this to be true. The maxilla appears to be normal in volume but malpositioned secondary to the abnormal CFM mandible.

4.2.4 Ear

As with all other manifestations of CFM, ear anomalies range from mild hypoplasia to complete anotia. From the OMENS classification, E1 represents mild hypoplasia, often with cupping of the affected ear; E2 represents absent auditory canal and

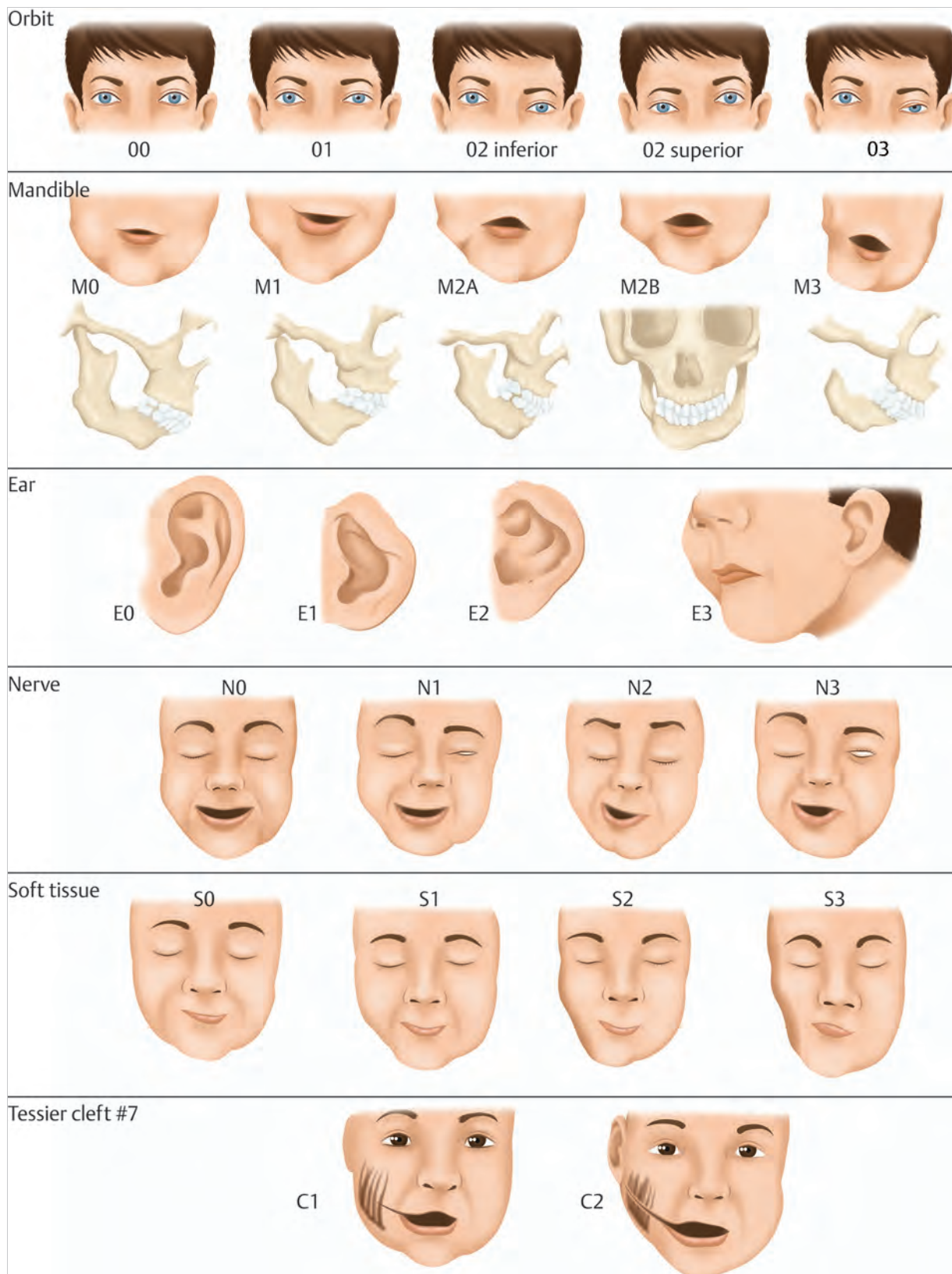


Fig. 4.1 OMENS classification. The severity of the clinical features of craniofacial microsomia is described in this classification. The specifics of each category can be found in their respective text in this chapter.



Fig. 4.2 Orbital deformity in craniofacial microsomia—microphthalmia. This figure demonstrates the severity of the deformity that may occur in the orbit. This patient had been previously treated for craniostylosis.

hypoplastic abnormal appearing ear remnant, and E3 represents severe microtia with all but a small remnant missing (e.g., lobular type). Preauricular remnants are also commonly found anterior to the tragus. Studies demonstrate that the ear is affected in 80 to 90% of patients, with as many as 50% of patients having microtia (E3). Hearing impairment is common on the affected side in CFM, occurring in approximately 75% of patients. Nearly all of those patients have conductive hearing loss; however, about 10% of all patients with CFM have sensorineural hearing loss as well.

4.2.5 Nerves

Anomalies of the nervous system, including aberrant brain anatomy, may be seen in CFM. However, the most common finding is partial facial nerve (CN VII) palsy. Facial nerve involvement is found in approximately 40% of patients with CFM. This palsy is more likely to affect the lower face (buccal, marginal mandibular, and cervical branches) than the upper face (zygomatic and frontal branches). A small percentage of patients with CFM have complete facial palsy. The OMENS classification describes facial palsy as either upper (N1), lower (N2), or total (N3).

Clinical manifestations of facial paralysis include inability to close the eyelid, lack of animation of the forehead, lack of active/symmetric smile, drooling, and asymmetry of lip depressors.

4.2.6 Soft Tissue

All layers of the soft tissue on the affected side of a patient with CFM may be deficient. This includes skin, subcutaneous fat, and muscles of animation and mastication. In moderate and severe cases, this deficiency immediately draws the attention of an observer to the asymmetry of the face. The most notable areas of deficiency are in the temporal fossa, in the malar/zygomatic region, and along the lower mandibular border. The severely affected CFM mandible contributes to the appearance of soft tissue deficiency, because the condyle and ramus are shifted medially, robbing the face of transverse width. The OMENS classification describes this soft deficiency as mild (S1), moderate (S2), or severe (S3). Macrostomia, or clefting at the oral commissure, also known as a Tessier 7 cleft, occurs in approximately 25% of patients with CFM. The orbicularis oris is discontinuous and can contribute to poor oral competence and abnormal appearance in both repose and animation.

4.2.7 Noncraniofacial Manifestations

Noncraniofacial anomalies were long thought to identify a patient with “syndromic CFM,” usually Goldenhar’s syndrome. The current understanding is that the error in neural crest cell migration responsible for facial manifestation of CFM is also the cause of noncraniofacial problems. The most common extracraniofacial anomalies occur in the heart (25% of patients with CFM), kidneys (10% of patients with CFM), skeleton (40% of patients with CFM), and central nervous system (CNS). Common cardiac defects include atrial and ventricular septal defects. Skeletal anomalies are most often scoliosis and overt vertebral abnormalities. The most common CNS problem is hydrocephalus. One of the pathognomonic findings is epibulbar dermoids of the eyelid.

4.2.8 Differential Diagnosis

A number of different diagnoses produce facial asymmetry that could resemble CFM to the untrained eye. Often, these other diagnoses can be excluded based on their timing of onset and specific phenotype. Examples of these diagnoses include Parry-Romberg disease, TMJ ankylosis, and radiation-induced tissue atrophy. Severe Tessier 7 can mimic CFM as well.

Facial asymmetry may be caused by hypoplasia or hyperplasia. Hemifacial hypertrophy and an infiltrating lipomatosis are rare conditions that create facial asymmetry. Condylar hypoplasia and hyperplasia should also be considered.

Treacher Collins syndrome (TCS) may be confused for bilateral CFM. Treacher Collins syndrome is a heritable (autosomal dominant) syndrome characterized by panfacial bone hypoplasia, bilateral Tessier clefts, and severe antimongoloid palpebral slant, culminating in a very characteristic phenotype.

One recently identified diagnosis, condylar-coronoid collapse, is believed to mimic CFM in producing mandibular conformation similar to mild to moderate CFM. It is noted to have a deep sigmoid notch, but the patients demonstrate no other OMENS-related features of CFM.

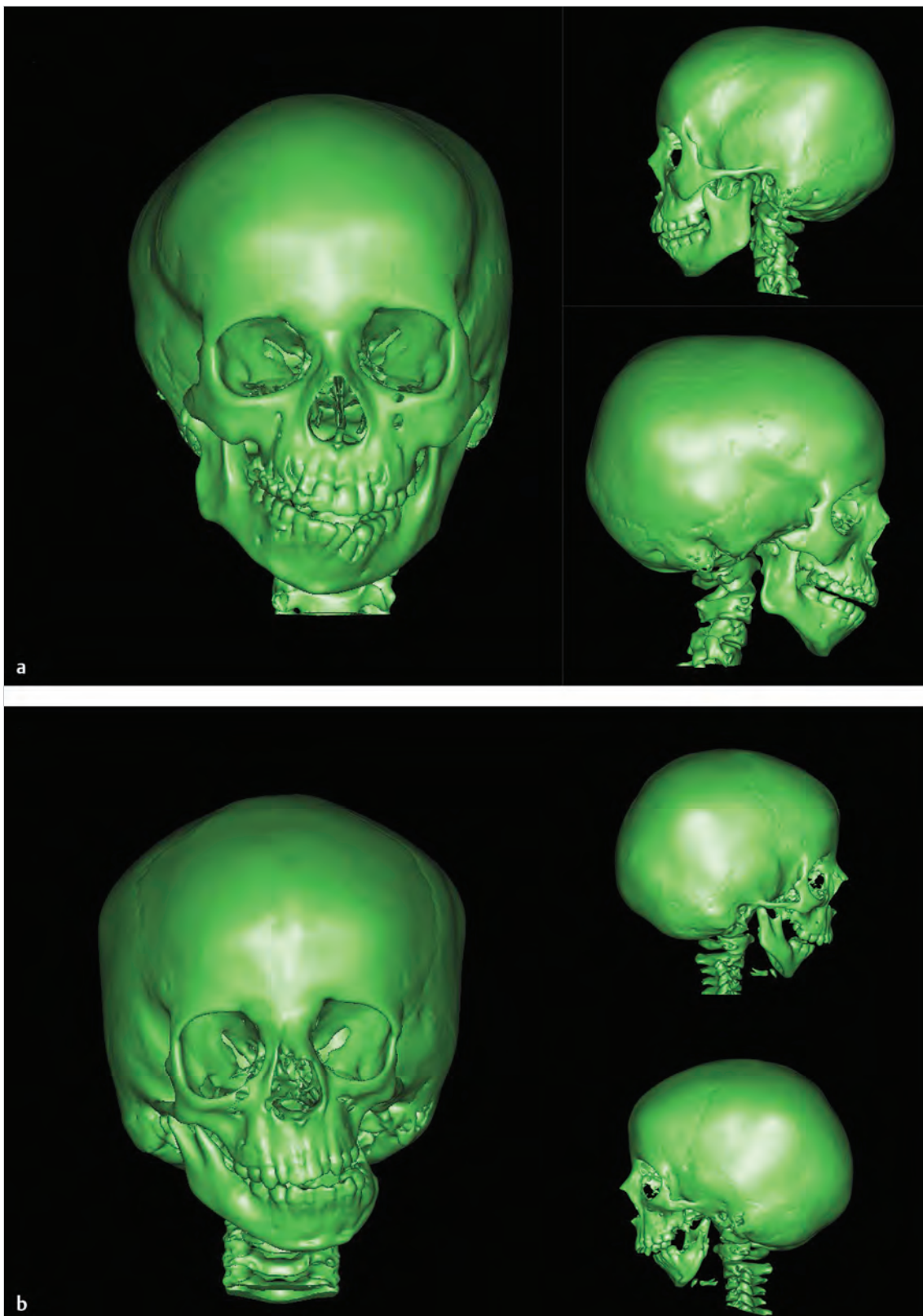


Fig. 4.3 Spectrum of mandibular deformities in craniofacial microsomia on CT scan. **(a)** CT demonstrates a smaller right mandibular ramus with an occlusal cant. The effect on the mandible is relatively mild. **(b)** CT demonstrates highly abnormal left hemimandible with absent ramus, condyle, and coronoid.

4.2.9 Natural History

The natural history of CFM is poorly understood. One fundamental question is whether the CFM deformity will worsen as a child ages. Furthermore, will interventions such as mandibular reconstruction mitigate or exacerbate age-related change? This has been studied most intensively pertaining to the mandible, which can be measured radiographically. Through a number of different methods, the affected mandible has been shown to grow but at a rate less than the rate of growth of the unaffected side. Given this, the phenotype of a growing patient is likely to worsen slightly as he or she grows (i.e., occlusal cant and facial soft tissue asymmetry).

4.2.10 Evaluation

Multidisciplinary craniofacial team care is critical in patients with CFM. This ensures both that the patients will be treated in an institution with the expertise and experience to handle such a complex constellation of abnormalities and that it will be done in a coordinated way. The team should include a geneticist, audiologist, speech pathologist, otolaryngologist, orthodontist, ophthalmologist, and craniofacial plastic surgeon.

4.2.11 Workup

Patients with CFM often receive a comprehensive workup. Extracraniofacial anomalies may necessitate a cardiac, skeletal, renal, and cerebral workup with the appropriate imaging modality and specialist consultation. In patients with significant mandibular deformity, visible retrognathia, or any observed breathing difficulty, a formal sleep study (polysomnogram) should be obtained to evaluate for obstructive sleep apnea. A formal endoscopic examination of the upper and lower airways may also be valuable if concerns arise about airway obstruction. Evaluation of cranial anatomy is most often done using three-

dimensional computed tomographic (3D CT) scan, which allows detailed examination of the entire craniofacial skeleton.

4.3 Operative Treatment

Surgical management of patients with CFM may begin immediately after birth and is likely to continue until skeletal maturity. Indications for, and timing of, surgery are not absolute, and the decision for reconstruction must be tailored to each individual's needs and desires. ▶ Fig. 4.4 demonstrates the treatment algorithm of the Children's Hospital of Philadelphia for patients with CFM.

4.3.1 Algorithm

Airway and nutrition are the top priorities when a new baby with CFM is born. Mild cases of CFM may go unnoticed in the newborn nursery, whereas severe cases may necessitate emergent intubation for airway compromise. Neonatal management of the impaired CFM airway may include endoscopic airway examination to rule out other anomalies such as choanal atresia and tracheomalacia. Endoscopy may also reveal or rule out tongue-based upper airway obstruction (TBUAO). In severe cases of bilateral CFM, it may be necessary to place a tracheostomy, especially if there is not enough bone stock for consideration of mandibular DO (MDO). In cases of TBUAO with sufficient bone stock, MDO may provide an alternative to tracheostomy. It is also important in patients with CFM to ensure proper nutrition—this may include nasogastric or surgically placed gastrostomy tubes. The algorithm for the treatment of all anomalies of CFM throughout the life of a patient is complex and controversial. ▶ Fig. 4.4 attempts to condense this treatment as a timeline. It is worth noting that many steps overlap, and this is an important consideration for the psychosocial well-being of patients with CFM.

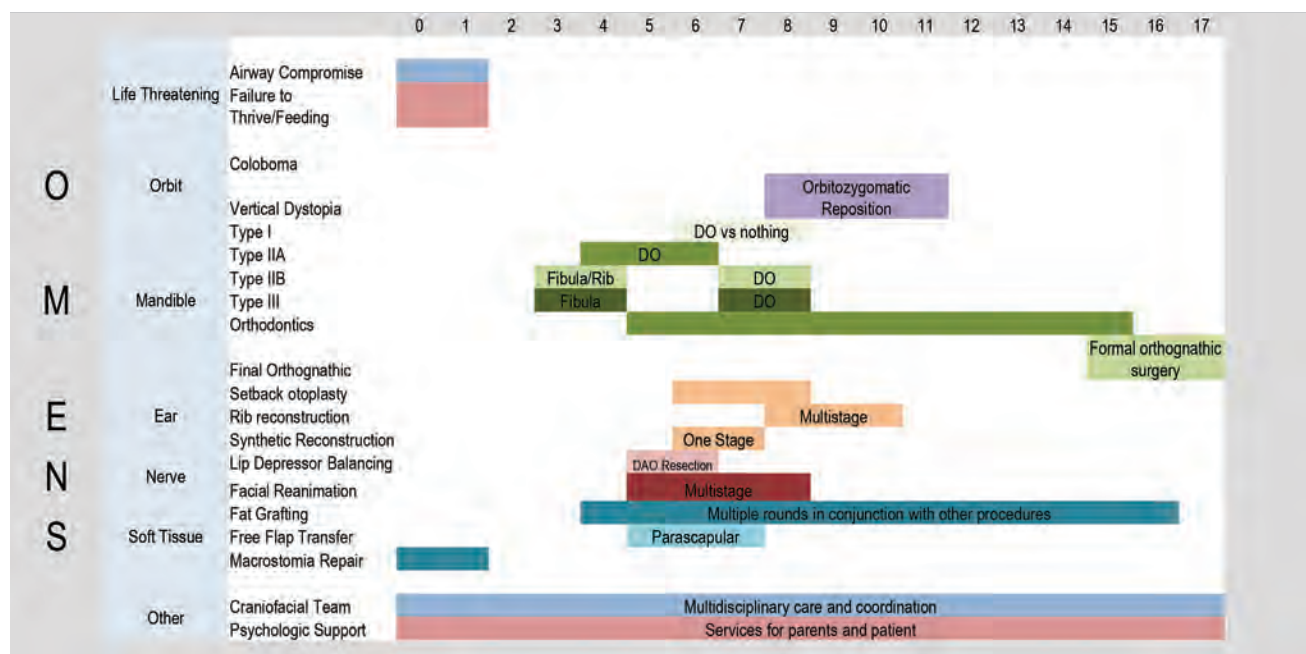


Fig. 4.4 Timing of treatment in craniofacial microsomia (CFM). This flow sheet represents the age at which the various CFM abnormalities are treated. DO, distraction osteogenesis; DAO, depressor anguli oris.

4.3.2 Macrostomia Repair/Commissuroplasty

Much like cleft lip repair, repair of the clefted oral commissure (macrostomia) can be performed in early infancy. This repair involves repositioning the commissure to an anatomic position that mirrors the contralateral side. This position is typically identified by observing the transition from cleft to noncleft tissue as well as by measuring commissure distances on the unaffected side (in unilateral cases). The principles of macrostomia repair are (1) repair of the clefted orbicularis oris muscle, (2) repositioning of the commissure to an anatomic and symmetric position, and (3) cutaneous scar rearrangement (i.e., Z-plasty) to avoid scar contracture and lateral drift of the commissure.

4.3.3 Orbital Reconstruction

Periorbital reconstruction includes the correction of colobomas as well as extraocular muscle imbalance (amblyopia) and is often done in consultation with an ophthalmologist. Correction of the orbit itself for vertical dystopia or severe constriction is much less common. Discrepancies in vertical height, if significant, can be corrected via cranial bone graft or alloplastic implant. The severely constricted orbit with microphthalmia may be treated with an orbital expander to allow for lid expansion and adequate space for a prosthesis. In cases where these techniques fail, it may be necessary to surgically expand the orbit, based on its anatomy—this can include zygomatic and/or supraorbital repositioning.

4.3.4 Mandible Reconstruction

The techniques for reconstruction of the CFM mandible can be divided into the following groups: DO, importation of

additional bone (graft and flap), correction of TMJ ankylosis, and final conventional orthognathic surgery at maturity. The goals of treatment are to level the occlusal plane and create bony mandibular symmetry. Distraction osteogenesis of the hypoplastic mandible is currently the most popular form of reconstruction, particularly during the period of mixed dentition. It relies on the principles popularized by Ilizarov for lower extremity lengthening after osteotomy and gradual distraction of the bony segments, with concomitant bone formation in the resultant gap.

Costochondral grafting is the most popular technique for TMJ reconstruction in CFM. Although some reports have highlighted the unpredictability of costochondral grafts, others have documented reliable symmetric growth in the majority of pediatric patients requiring TMJ reconstruction for CFM. Free tissue transfer in the form of a free fibula may be required when condyle/ramus and body must be reconstructed. In general, less severely affected patients can be treated at a later age and with fewer surgical stages. The Kaban-modified Pruzansky's classification provides a conceptual framework for understanding the timing and type of treatment.

4.3.5 Treatment by Subtype

Patients with type I deformity are often successfully treated with orthodontia or single-stage orthognathic surgery at skeletal maturity. Distraction osteogenesis in early permanent dentition may be offered when a significant dental cant exists, but this is an exception rather than a rule. The decision to avoid or delay mandibular procedures until skeletal maturity is often based on the degree of psychosocial distress that is experienced by these patients. In patients with minimal deformity, anxiety and stress associated with facial asymmetry can usually be mitigated or temporized without major mandibular procedures (► Fig. 4.5).

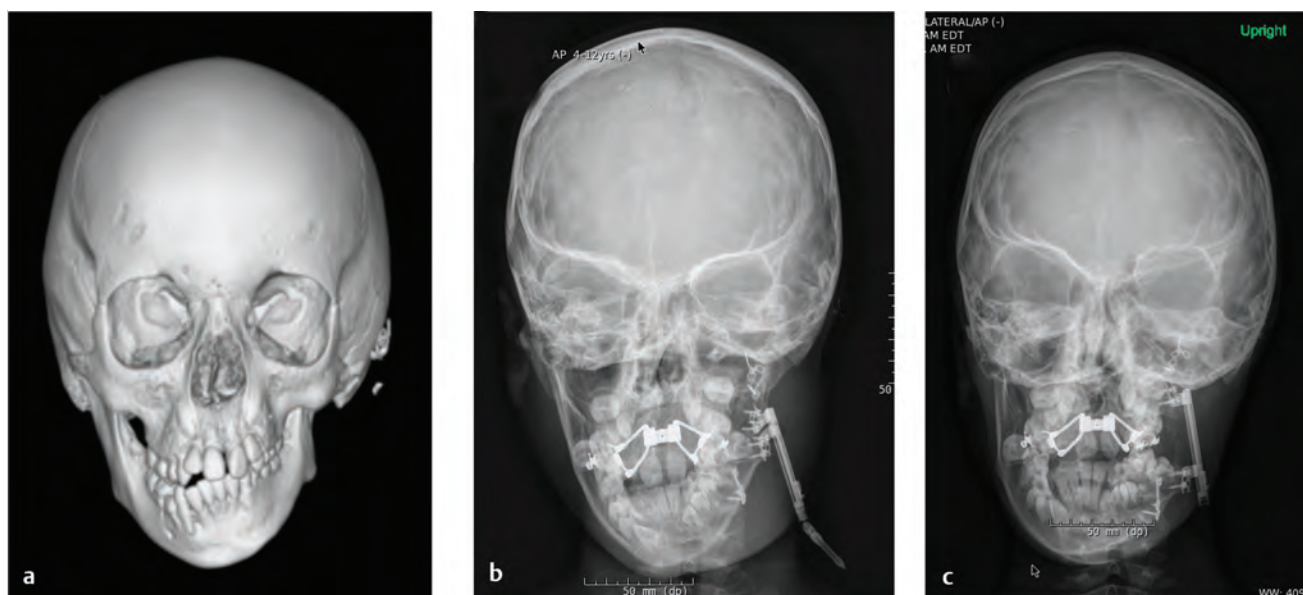


Fig. 4.5 Vertical mandibular distraction for vertical mandible. (a) 3D CT scan demonstrating small left mandible with occlusal cant and vertically shortened ipsilateral maxilla. (b) Distractor in place but before distraction. (c) Full distraction of 20 mm with orthodontic closure of open bite during distraction activation phase.

Distraction osteogenesis with or without rib graft reconstruction is the most common treatment modality for type II mandibular hypoplasia. Unlike type I mandibular hypoplasia, the facial asymmetry in type II patients is often significant enough to result in functional problems with occlusion as well as in anxiety and psychosocial distress. Surgical intervention can provide both functional and aesthetic improvements, at least in the short-term follow-up. Type IIa mandibular hypoplasia can often be addressed with mandibular DO alone, whereas the abnormal TMJ in type IIb mandibular hypoplasia may necessitate costochondral grafting, with or without subsequent distraction.

Similar to type IIb mandibular hypoplasia, type III deformity may benefit from surgical intervention during mixed dentition, thereby mitigating the functional and aesthetic concerns associated with such severe deformities, at least during the formative childhood and early adolescent years. Costochondral grafting with or without mandibular DO is the most common form of treatment. As stated previously, growth of the costochondral graft may be unpredictable, with some reports describing lack of growth, whereas other reports have documented overgrowth. Several centers have reported satisfactory results with osteocutaneous free tissue transfer to treat severe type IIB and III mandibles, as well.

It is critical to recognize that all patients with CFM may benefit from finishing conventional orthognathic surgery at skeletal maturity. This will be technically more challenging because of the magnitude of the deformity, the presence of scar, and the anatomic abnormality of the reconstructed mandible. The use of computer-based presurgical planning can be invaluable in these situations (► Fig. 4.6 and ► Fig. 4.7).

4.3.6 Ear Reconstruction

Ear reconstruction must compete with all other reconstructive needs in patients with CFM. The timing of ear reconstruction is typically after jaw correction and before hearing restoration (auditory canaloplasty or bone-anchored hearing aid placement). Ear reconstruction would ideally be performed at a young age, before a child's peers have developed cruel tendencies and before significant psychological harm has been done. The challenge of early ear reconstruction is that the template for this reconstruction, namely the contralateral ear, has not yet reached adult size and the building material for a new ear, namely rib cartilage, is not large or firm enough to create an anatomic ear. Because of these competing interests, rib cartilage-based ear reconstruction is often done at around 8 to 10 years of age (► Fig. 4.7). Many experienced surgeons have noted disappointment and unpredictability of carved rib cartilage ear reconstructions. In addition, the donor site can be quite painful and morbid. An alternative to this is a synthetic ear construct (porous polyethylene). These ear implants can be performed earlier in life at around 5 to 6 years of age. Use of a synthetic ear reconstruction requires coverage with a temporoparietal fascial flap to cover the construct in its entirety. This flap is then covered with full-thickness skin grafts. There is now high-quality long-term evidence that these synthetic ears are durable.

Additional alternative include prosthetic ears, which are attached with either adhesive or bone anchored magnets. Insurance often does not cover prosthetic reconstruction, which can cost in excess of \$8,000 and have a limited life span.

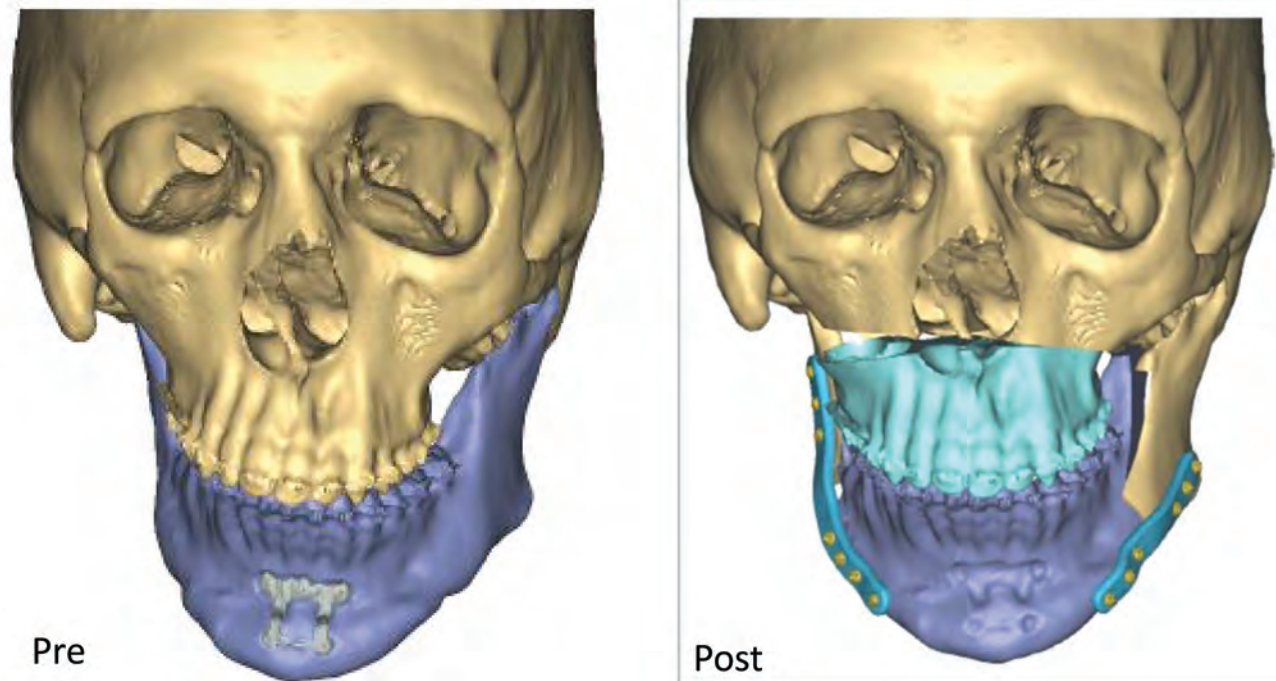


Fig. 4.6 Computer-aided surgical planning of final orthognathic surgery. This patient with craniofacial microsomia (CFM) underwent mandibular distraction osteogenesis once in childhood and presents at skeletal maturity for correction of recurrent occlusal cant and facial asymmetry. Final orthognathic surgery in CFM necessitates complex, large-magnitude movements. 3D computer planning can make this process much more efficient. Of note, in this case, the patient had previously had a genioplasty to mask asymmetry, and the operated/abnormal CFM mandible may require rigid fixation, as seen here.



Fig. 4.7 Female with right craniofacial microsomia who has undergone one round of unilateral mandibular distraction in childhood, multiple rounds of fat grafting, rib-based ear reconstruction at an outside institution, and final two-jaw orthognathic surgery at skeletal maturity. **(a,b)** 9 years: Predistraction with evident occlusal cant and facial asymmetry. **(c,d)** 13 years: Preorthognathic surgery. **(e,f)** 17 years: Post-Le Fort I, bilateral sagittal split osteotomy, and advancement genioplasty.

4.3.7 Facial Reanimation

Reanimation of the paralyzed face is a technically demanding field. The patient and the surgeon must strike a balance between many important factors, including realistic expectations, volitional versus natural animation, one stage versus multiple stages, and no donor site versus multiple donor sites. Early in life, critical attention must be paid to corneal protection. If there is concern for lagophthalmos, the inability to close the eye completely, then a regimen of frequent lubrication must be started. Temporary tarsorrhaphy may also be performed. In general, consistent lid closure is obtained with the insertion of a gold weight into the upper eyelid. However, in neonates, infants, and small children, the eye must be closely monitored for exposure keratitis.

Another goal of facial reanimation is to improve social interactions and acceptance by creating an animated smile. In children, this can be accomplished by means of mimicking a smile when a nerve or muscle of mastication is used. Alternatively, this can be accomplished spontaneously by powering animation from the contralateral functioning facial nerve. The most commonly used strategy is a two-stage facial reanimation, combining a sural nerve graft with neurotized free gracilis transfer to allow a spontaneous and coordinated smile. Similarly, the temporalis muscle can be transferred from its insertion on the coronoid process of the mandible to the modiolus (the Labbe's procedure).

4.3.8 Soft Tissue Augmentation

Patients with CFM are often deficient of subcutaneous facial volume. After the advent of microsurgical tissue transfer (free flaps), the most common choice for augmentation was the free parascapular flap (► Fig. 4.8). This flap was tailored to the morphology of the deficiency and placed via a preauricular incision with anastomosis to the facial vessels. This procedure would often be a finishing procedure, as any surgical treatment for mandibular asymmetry, facial palsy, and microtia should be performed first. In the age of microfat injection, with Coleman's technique, a new paradigm has arisen. In mature patients with severe soft tissue volume deficiency, there is still a role of one-stage free tissue transfer. However, fat grafting now offers the opportunity to slowly build facial volume, with minimal morbidity and downtime (► Fig. 4.7). In fact, facial fat grafting can be done at the time of nearly all other corrective procedures for CFM. Fat is harvested where available (abdomen, inner thigh, and buttocks) and tailored to the deficiency. On average, 10 to 25 mL can be placed, understanding that, at most, two thirds will be revascularized and retained. Fat grafting typically requires three to five or more stages (► Fig. 4.7 and ► Fig. 4.8).



Fig. 4.8 A male patient with severe right craniofacial microsomia and a grade III hemimandible. **(a,b)** Patient at the age of 7 years before any surgical intervention. **(c,d)** Patient at the age of 20 years after undergoing rib grafting, right free parascapular flap, formal orthognathic surgery with genioplasty, and multiple rounds of fat grafting.

4.4 Complications

4.4.1 Mandible Reconstruction

Complications related to mandible reconstruction include tooth bud injury, lip numbness, partial facial palsy, and malocclusion. Hardware can fail. Free flaps and rib grafts may fail via resorption, nonunion, or fracture. If present, the TMJ may become ankylosed or a condylar reconstruction may fuse to the temporal bone.

Many complications from mandible reconstruction are specific to the modality that is used. A critical consideration is to

anticipate that procedures performed in early childhood will likely be insufficient for the entirety of the child's growth period (i.e., skeletal maturity). With DO, early postoperative complications include wound infection/abscess and device failure. Technical complications include improper vector of distraction and partial facial palsy, when using a submandibular approach. Other important late complications include bony nonunion and relapse. Distraction of the mandible unilaterally can result in persistent malocclusion (unilateral open bite)—in children younger than 10 years, maxillary down growth is typically obtainable with orthodontics after the maxilla has been offloaded.

Complications related to large reconstruction depend on technique. Free bone flap transfer can be complicated by flap loss. A long-term complication from free fibula use can be lack of growth, which leads to future asymmetry. Another important complication can be ankylosis of the proximal condylar segment to the temporal bone; this necessitates a future TMJ release. Donor-site complications can occur at both free fibula harvest and a costal bone harvest.

4.4.2 Ear Reconstruction

In the short term, extrusion, infection, and exposure of either rib or synthetic ear constructs are the most concerning complications. Superficial wound infections can be treated with antibiotics, whereas deep space infections require operative debridement. Exposure of the ear construct is treated with operative washout; debridement of necrotic, infected tissue; and flap closure. Small extrusions of synthetic ear constructs have successfully been treated in this way—soft tissue coverage is often provided by an occipital fascial flap.

Long-term complications are largely related to aesthetics. Microtia reconstruction with rib cartilage is often accompanied by poor definition of the complex 3D details. This is related to scar formation. Synthetic ear constructs are thought to have enhanced and improving definition over time. However, there have been reports of late synthetic ear extrusion necessitating removal. The other important aesthetic complications to note are malposition and size mismatch.

4.4.3 Facial Reanimation

Complications of facial reanimation surgery include loss of muscle flaps, injury to other neurovascular structures during facial dissection, and dehiscence of inset muscle. It is a multi-staged operation, with complete failure as a possibility at any stage. In the end, it may produce underwhelming results, with the lack of a symmetric, spontaneous smile. In addition, although poor animation of the smile is not a complication per se, it may be seen as such by patients and parents. This highlights the importance of preoperative counseling and creation of realistic expectations.

4.4.4 Soft Tissue Augmentation

For free parascapular flap reconstruction (► Fig. 4.8), complete or partial flap loss is a known complication that occurs in approximately 1 to 5% of patients in high-quality series. Partial flap loss can present as a loss in volume as well as hardened, calcified areas of fat necrosis or oil cysts. Coleman's fat grafting should retain approximately two thirds of the delivered volume in the long term. These grafts too can be complicated by oil cyst formation and poor take with little volume retention. Donor-site complications can occur in both flaps and fat graft harvest.

4.5 Conclusion

Craniofacial microsomnia represents a significant reconstructive challenge for plastic craniofacial surgeons. Each of the OMENS categories can represent a significant surgical undertaking, and taken as a whole, it represents many surgeries throughout the life of a patient with CFM.

The authors would like to acknowledge the contributions of Scott P. Bartlett, MD, to this chapter.

4.6 Key Points

Craniofacial microsomnia represents an embryologic error in neural crest cell migration, affecting the first and second branchial arches primarily.

- Its clinical manifestations are succinctly organized in the OMENS-Plus classification system—orbit, mandible, ear, (facial) nerve, and facial soft tissue hypoplasia, plus assorted noncraniofacial anomalies.
- The spectrum of presentation is highly variable, from mild facial asymmetry to severe presentations with microtia, hypoplastic mandible, obvious facial asymmetry, and facial paralysis.
- Surgical treatment is customized to the severity of the deformity and to the anatomic region—multiple stages, relapse, and surgical treatment from early childhood to skeletal maturity are the rule.
- Distraction osteogenesis for the mandible and particulate fat grafting for soft tissue deficiency represent paradigm shifts in the management of these patients.

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5 Orbital Hypertelorism

Robin Yang, Srinivas M. Susarla, and Anand Kumar

Summary

Hypertelorism remains a clinical finding associated with a variety of congenital conditions. The severity of hypertelorism can be based on the interdacryon distance and can be used in conjunction with the shape of the frontal-orbital region to better characterize the orbital dysmorphology. The correction of hypertelorism is not a life saving procedure and is mainly an aesthetic procedure when intracranial pressure is normal. Thus, a careful risk benefit analysis will be required for each case with the treatment plan tailored to each specific deformity and patient related variable. The appropriate choice depends on the relationship between the orbits and the surrounding craniofacial structures:

If the maxillary arch is narrow and the corresponding dentition shows apertognathia with the incisors more cranial than the molars, a bipartition is the operation of choice to improve the maxillary arch and the orbits in the same procedure.

If a unidirectional repositioning is needed in less severe (IOD < 35 mm) cases, a box osteotomy is satisfactory. If the orbits are asymmetric with laterally and downwardly oblique positioning, a bipartition is preferred.

Bipartition is the procedure of choice in the most severe cases.

If the nasal structures are narrow, a bipartition and medialization of the upper face can improve the airways by enlargement of the lower face.

An encephalocele associated with a midline cleft can easily be accessed by a bipartition or orbital box procedure. The amount of periobital and midface well can be quite significant. Without tracheostomy, we prefer 72 hours of post surgical intubation and sedation. This allows maximal swelling to occur and begin resolution and ensures adequate hemostasis prior to extubation. Early extubation has led to serious mal-outcomes in our experience and thus our abandonment of this practice. When the patient meets clinical parameters for extubation, it should be performed within 5 days of surgery to assist with patient mobilization and enteral feeding. Blindness, diplopia, and anosmia are uncommon complications, but should be discussed with the patients.

Keywords: telorbitism, midline cleft defect, Tessier 0/14 cleft, horizontal dystopia

5.1 Introduction

Orbital dystopia can be defined as the abnormal displacement of the orbit and its contents in relation to the adjacent craniofacial skeleton or more specifically in the vertical and/or horizontal planes. Orbital dystopia in the horizontal plane has been termed orbital hypertelorism. Hypertelorism is a symptom or associated finding that can be accompanied by a multitude of craniofacial conditions that are not genetically or epigenetically related. Tessier had classically stressed the fact that hypertelorism is the displacement of the orbital cones in their entirety,

manifested by expansion of the central ethmoid sinuses and widening of the sphenoid wing angle to more than 110 degrees. The severity of hypertelorism can be based on the interdacryon distance and can be used in conjunction with the shape of the frontal-orbital region to better characterize the orbital dysmorphology. Patients presenting with orbital hypertelorism will require multidisciplinary specialty evaluation and coordinated care. The specialties include but are not limited to ophthalmology, oculoplastic surgery, psychology, social work, pediatrics, neurosurgery, pediatric dentistry, craniofacial orthodontics, and craniofacial surgery.

5.2 Diagnosis

Orbital hypertelorism is the physical manifestation of excessive and widened ethmoidal sinuses. This orbital anomaly is not pathognomonic of any particular condition but can present in a variety of conditions, including craniofacial clefts, craniofacial dysplasia, and facial craniosynostosis syndromes. Although the term *hypertelorism* is frequently misused to describe an increased intercanthal distance following trauma or tumors, this process does not cause a change in position of the lateral wall of the orbit or sphenoid wing angle. Hence, most causes of traumatic increase in intercanthal distance are properly termed *telecanthus*.

The distance between the two medial canthi is known as the intercanthal distance. The distance between the two lateral canthi is known as the outer canthal distance. Interpupillary distance is the space between the two midpupillary points in forward gaze (► Fig. 5.1). All three distances (interpupillary distance, intercanthal distance, and outer canthal distance) are increased in patients with hypertelorism. In patients who suffer from a trauma (nasoorbito-ethmoid fractures), there is usually an increase only in the intercanthal distance, resulting in *telecanthus*.

In the preoperative setting, a computed tomographic (CT) scan is recommended as the first-line diagnostic tool. This can detect any previous or ongoing congenital brain malformations. Any malformations identified should be treated by a neurosurgeon in a collaborative and, often, staged reconstructive strategy to optimize outcome. A high-resolution CT scan with thin 1-mm slices will also aid in accurate three-dimensional (3D) reconstruction by limiting the effects of volume averaging of the craniofacial skeleton. In many cases, it can also reveal where certain vital structures are located (e.g., maxillary tooth buds if a Le Fort osteotomy is planned in addition to the orbital repair).

Patients with orbital dystopia may present with a variety of anatomical differences in the anterior cranial base. The ethmoid sinuses, nasal cavity, and sphenoid wings are usually abnormally lengthened. The crista galli and cribriform plates are usually displaced inferiorly, especially in patients with facial clefts. These anatomical abnormalities can cause defects in the anterior cranial base. Appropriate preoperative planning can avoid surgical complications and subcranial strategies such as the Le Fort III osteotomy are the best surgical options. Virtual surgical planning (VSP) has become the de facto standard of care

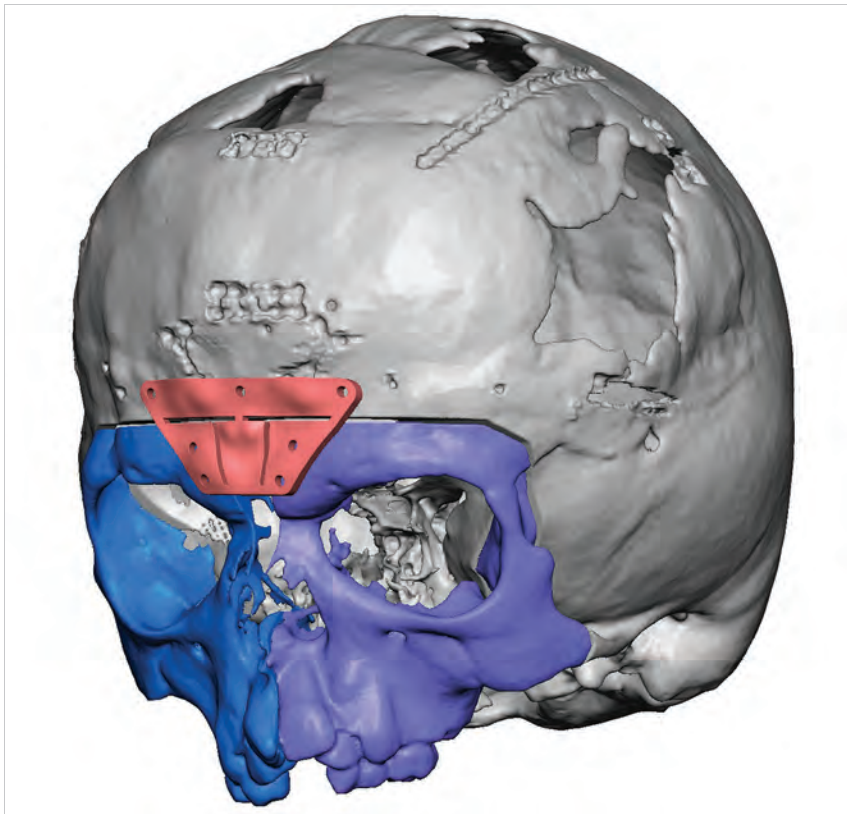


Fig. 5.1 The facial bipartition can be easily designed by using virtual surgical planning. This oblique image demonstrates the segmentation of face and central wedge resection in a patient with Apert's syndrome. The central face concavity can be readily corrected by bowing the face anteriorly after bipartition. The cutting guide in place aids in the precise marking of the central osteotomy.

regarding osteotomy design and planning. The cost of VSP has largely been absorbed into the cost of the craniofacial fixation system or distraction system, thus facilitating more widespread adoption. Similar in process to VSP for orthognathic surgery, a more commonly performed procedure, VSP for orbital deformities has added in accurate measurement of the interdacryon distance and thus measurement of the central segment of bone required for optimal correction of the orbits (► Fig. 5.1). We prefer to use an interdacryon distance of 18 mm as the final distance and split the difference equally in symmetric cases or one third to two thirds in asymmetric cases, commonly seen in the X-linked cranial frontal nasal dysplasia syndrome.

It is also important to have the patient evaluated by an orthodontist who is preferably trained for craniofacial deformities. Many craniofacial teams have appropriately trained orthodontists who are comfortable with the management of the difficult occlusion that may present in a patient with hypertelorism as a sequel of his or her craniofacial deformity. Frequently, abnormal position of the orbital cones can directly impact the maxilla. Apertognathia is often seen in these patients, because the smaller ethmoid bones limit the normal caudal rotation of the maxillary ridge in development. In addition, a constricted maxilla with a high arched palate can often be encountered. We prefer the use of the facial bipartition to address the constricted maxillary arch when both orbital constriction and maxillary expansion are needed. In contrast, the orbital box osteotomy is more optimal when only orbital expansion needs correction. Facial advancement and the correction of sleep apnea along with orbital pathology can be accomplished only with the facial bipartition, thus making this operation our preferred and most versatile corrective procedure.

5.2.1 Nonoperative Treatment

The correction of hypertelorism is not a life-saving procedure and is mainly an aesthetic procedure when intracranial pressure is normal. Thus, a careful risk-benefit analysis will be required for each case, with the treatment plan tailored to each specific deformity and patient-related variable. Patients with severe noncorrected congenital heart disease or severe developmental delay who are noncommunicative or vegetative are not considered candidates for surgery because of the unfavorable risk-to-benefit ratio.

5.2.2 Operative Treatment

Corrective surgical procedure for hypertelorism is primarily indicated when the deformity is thought to cause significant facial distortion, resulting in significant poor self-esteem and body image and is in conjunction with a comprehensive approach to the craniofacial skeleton. The upper face and orbits need to be set in their proper position before traditional orthognathic surgery in most patients for optimal restoration of facial balance and harmony. Many patients will have normal mental and physical development despite their often-disfiguring appearances, and this remains a major reason for comprehensive surgical correction of hypertelorism. The goals of surgical corrections of hypertelorism, regardless of cause, should be to:

- Normalize the anatomical distance between the orbital cones.
- Narrow the nasal width and create an appropriate nasal projection.
- Correct any soft diffuse deformities or clefts as well as normalize hair-bearing areas (widow's peaks or eyebrows).

5.2.3 Operative Timing

All pediatric craniofacial reconstructions are a fine balance between managing appropriate growth of the craniofacial skeleton and optimizing psychosocial development and societal integration. The advantage of operating earlier in life is to address the psychosocial aspects as the developing child becomes more self-aware. The best and most stable results for correction of hypertelorism are obtained after the growth of the cranium and upper one-third of the face or orbits has completed. By the age of 6 years, the majority of the interzygomatic width is established and there is adequate descent of the maxillary tooth buds to facilitate an osteotomy below the infraorbital nerve. Before the age of 5 years, the orbital bones are thin and fragile, which can make the operation more difficult and affect the stability of the fixation, leading to relapse. Most craniofacial centers proceed with hypertelorism correction between the ages of 6 and 8 years. Our preferred age of treatment is 8 years, a time when the upper third of the facial skeleton has completed the growth, but before the development of the frontal sinus, which is typically seen at the age of 10 years.

5.2.4 Operative Selection

Before the landmark work of Paul Tessier in the late 1960s, correction of hypertelorism was not possible. The combined intra- and extracranial technique allowed appropriate repositioning of the entire orbital cones. The gold standard of correction is a choice between orbital shift using either box osteotomies or facial bipartition. The appropriate choice depends on the relationship between the orbits and the surrounding craniofacial structures:

- If the maxillary arch is narrow and the corresponding dentition shows apertognathia, with the incisors more cranial than the molars; a bipartition is the operation of choice to improve the maxillary arch and the orbits in the same procedure (► Fig. 5.2).
- If a unidirectional repositioning is needed in less-severe ($IOD < 35$ mm) cases, a box osteotomy is satisfactory. If the orbits are asymmetric, with laterally and downwardly oblique positioning, a bipartition is preferred.
- Bipartition is the procedure of choice in the most severe cases.
- If the nasal structures are narrow, a bipartition and medialization of the upper face can improve the airways by enlargement of the lower face.
- An encephalocele associated with a midline cleft can easily be accessed by a bipartition or orbital box procedure.

5.2.5 Orbital Box Osteotomy

The orbital box osteotomy allows for mobilization of the entire orbital cones into a more normalized position. This procedure is routinely used to correct vertical orbital dystopia. However, as stated earlier, some craniofacial surgeons still use this procedure in patients with hypertelorism and a normal occlusion. Most craniofacial teams prefer the frontal approach, which allows the surgeon to have an unobstructed view of the anterior cranial fossa. This exposure also allows the neurosurgeon to repair any dural tears that are recognized. The design of the frontal craniectomy must be carefully planned. Surgical preference usually dictates the lower limit of the frontal craniectomy.

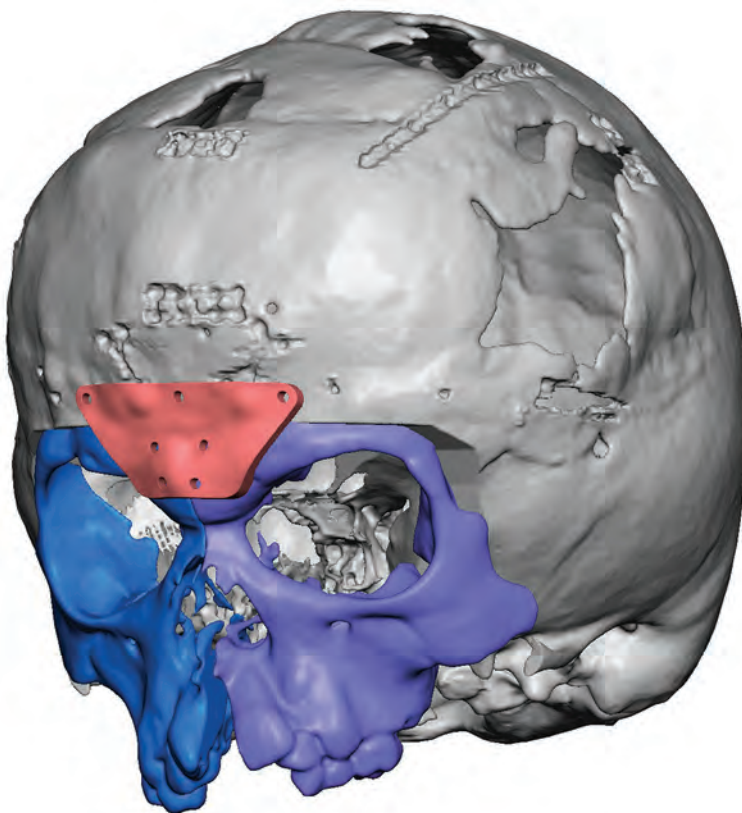


Fig. 5.2 After facial bipartition and central bone resection, a fixation guide can be used to mark optimal orbital translocation and temporary fixation by using a wire before plate and screw fixation. Note the large central gap between the central incisors after proper central orbital translocation.

Some surgeons prefer to keep a frontal bandeau intact at the lower part of the forehead. Usually, a frontal bandeau of approximately 1 cm is maintained between the frontal bone flap and the osteotomies of the supraorbital rim. The supraorbital rim to be preserved is also usually 1 cm in height, and the lower limit of the frontal craniotomy should be 2 cm above the orbits. Most craniofacial surgeons do not preserve the horizontal bandeau that was originally described by Tessier since the advent of rigid internal fixation. The horizontal bone segment was typically used to anchor wire fixation of the mobile bone segments. The lower limit of the craniotomy is kept 1 cm above the orbits. The frontal craniotomy approach allows access to the anterior cranial fossa.

5.2.6 Operative Details

Patient Positioning and Preparation

An oral endotracheal tube is placed and secured to the lower central incisor teeth by using a 24-gauge wire. The upper and lower eyelids can be closed with a tarsorrhaphy suture and placement of ointment to protect the cornea. A small strip of hair is trimmed and saved along the planned coronal incision. The authors' choice of local anesthesia is 0.5% lidocaine with 1:100,000 epinephrine. This is injected into the planned coronal incision, the maxillary vestibule, and adjacent to the infraorbital and supraorbital nerves. The patient is placed in neck extension into a standard headrest, where the entire head, face, and ears are prepped and exposed into the surgical field.

Surgical Exposure

A subperiosteal coronal incision is performed, and the dissection is carried out over the supraorbital rims. Using a small 2-mm osteotome, a small osteotomy is usually performed to carefully release supraorbital nerve from its foramen. The temporalis muscle is left attached to the cranium, and dissection is performed deep to the temporoparietal fascia, until the zygomatic arch is encountered. An incision is performed onto the zygomatic bone at this level to prevent fat atrophy and future temporal hollowing. The lateral rim attachment of the temporalis muscle is released to provide access for the bur holes needed for craniotomy as well as the inferior oblique fissure. A careful dissection of the orbital cavity and the nasal processes of the frontal bone are performed carefully to not damage the optic nerve. The periorbital is released circumferentially along the orbital cavity. The anterior ethmoid arteries serve as an anatomical landmark and are identified. Meticulous dissection is performed around the medial canthal tendon attachment as well as the nasolacrimal apparatus to avoid detachment or injury to either structure. Appropriate surgical exposure will include bilateral zygomatic arches, superior orbital rims, superior aspect of the nasal processes of the frontal bone, medial wall of the orbit, and the lateral orbital rim. An upper maxillary sulcus incision is performed to expose the nasal aperture, pyriform rims, zygomatic-maxillary buttresses, and inferior orbital rims. The infraorbital nerves should be identified to avoid injury, but care should be taken to minimize soft tissue stripping of the midface.

Frontal Craniotomy

It should be emphasized that in this patient population, anomalies are frequently encountered (thick or bifid crista galli and deep groove for the longitudinal sinus). The craniotomy bur holes are typically made in the pterional region. After the frontal flap is elevated, dissection of the dura from the orbital roofs, greater wing of the sphenoid, and the neighboring temporal fossa is performed. In cases of a large medial resection or if the olfactory grooves run close to the medial walls of the orbit, sacrifice of the olfactory nerves may be unavoidable. We prefer to place our medial osteotomy anterior to the crista galli to avoid injury to the olfactory system. A pericranial flap is useful to assist with nasal cranial separation at the completion of the osteotomies. A meticulous repair of any dura tears is mandatory. A dural patch using pericranium or deep temporal fascia is often used to reinforce the closure as needed.

Orbital Osteotomy

A reciprocating saw through a temporalis muscle tunnel is used to osteotomize the lateral orbital wall, starting at the inferior oblique fissure. Malleable retractors are used to protect the periorbital and its contents. Malleable retractors are also useful for retraction of the brain to expose the anterior central cranial base anterior to the crista galli. The supraorbital and upper third of the medial orbit osteotomies are performed transcranially. The medial thirds of the orbital floor are completed by using a 2-mm osteotome. These are performed at the centrum of the globe, typically 2 cm posterior to the orbital rim, to facilitate appropriate translocation of the globes. The zygomatic bones (including the zygomatic arch and its attachment to the maxilla) are osteotomized with a reciprocating saw, through the gingivobuccal and coronal incisions. The maxillary osteotomy is made typically 5 mm below the infraorbital nerve. The canine tooth bud remains the only major structure to avoid in the maxilla. Care to place the medial maxillary cut below the nasolacrimal duct is necessary to avoid postsurgery epiphora.

Anterior Cranial Base Osteotomy

A V- or U-shaped osteotomy of the nasofrontal region is based on the desired movement of the orbits and zygoma. The osteotomy is performed transcranially in the midline, extending anteriorly to the crista galli. The paranasal osteotomies are performed vertically, with a slight divergent angle. The frontal bone, ethmoid, and nasal septum are removed en bloc. Inspection of nasal perforations is also performed, and these are repaired.

Mobilization of the Orbits

Once all of the osteotomies are performed and hemostasis is achieved, the orbits can be mobilized. Smith spreaders can be used in a methodical fashion, starting at the lateral orbital rims and walking them forward. This allows the operator to visualize any areas that are not fully osteotomized. Attention should be paid to the pyriform rims as well as to any remaining nasal septum or posterior ethmoid cells that need to be removed for full mobilization. The orbits are then passively mobilized medially, and a 24-gauge wire is used for temporary anchorage. The

interorbital distance should be close to 18 mm and may account for 2 to 5 mm of overcorrection. Virtual surgical planning has assisted with accurate orbital movement in asymmetric cases; a contralateral normal side or a control normal side can be superimposed and used to accurately measure planned orbital movements (► Fig. 5.3 and ► Fig. 5.4).

Skeletal Fixation and Soft Tissue Draping/Closure

Rigid skeletal fixation is applied to the midline, zygoma, and pyriform regions. Often, the bone dust from the burr holes and bone fragments from the resections can be used to fill in any vault gaps. Cranial bone grafts are generally necessary to fill the gaps of larger defects as well as for nasal reconstruction. Bilateral medial canthopexy is performed, as needed, to augment the medial canthal tendon and the surrounding soft tissue. Typically, a soft tissue bolster is all that is needed if the medial canthus is left attached. The coronal incision is closed in a two-layer fashion. The oral mucosa can be closed with an absorbable interrupted suture.

5.2.7 Facial Bipartition

As discussed previously, timing of correction of hypertelorism is a balance of physiology and psychology. Generally, after 4 years of age, the skull is thick enough to perform stable fixation and mobilization of the segments. In skeletally mature patients, Kawamoto describes the use of a facial bipartition with a Le Fort I osteotomy. This allows control of the maxillary dentition and correction of any occlusal/skeletal discrepancies. In other clinical scenarios (e.g., Apert's syndrome), the bipartition can also be combined with a Le Fort III advancement as part of a secondary procedure following an initial anterior skull remodeling

performed at an earlier age. In general, the facial bipartition is the preferred procedure to correct orbital hypertelorism.

Patient positioning, preparation, exposure, and craniotomy are performed identical to the details discussed in the section on orbital box osteotomy. Of note, when performing the facial bipartition as an isolated procedure, small bilateral gingivobuccal incisions are made to facilitate the use of the pterygomaxillary osteotomies.

Osteotomies

Bilateral osteotomies are made anterior to the zygomatic arch, lateral orbital wall, orbital roof, medial orbital wall, pterygomaxillary buttress, and the nasal septum. The lateral orbital wall osteotomy is made with a reciprocating saw. The saw is passed through the temporalis muscle tunnel. The saw blade is oriented superiorly and directed toward the inferior orbital fissure. The osteotomy is carried up to the orbital roof and then across the anterior cranial base anterior to the cribriform plate at the midline. A small (4-mm) osteotome is used to create the "doughnut osteotomy" around the entire orbit. Areas of concern include the medial canthus and the lacrimal apparatus. Osteotomies should be made behind the medial canthal attachments. Intraoral use of the Kawamoto's osteotomes to make the pterygomaxillary osteotomies finalizes the craniofacial disjunction. Before down fracture of the monobloc, the intradacryon distance is measured. A V-shaped resection is planned to leave 16 to 20 mm of bone between the medial canthi region. The resected region includes the mid portion of the superior orbital rim, the radix, and the nasal bone. A small intraoral gingivobuccal incision is made to facilitate splitting of the palate.

The monobloc down fracture is performed with Rowe disimpaction forceps. A double-pronged nasal septal osteotome is also introduced and directed toward the junction of the hard

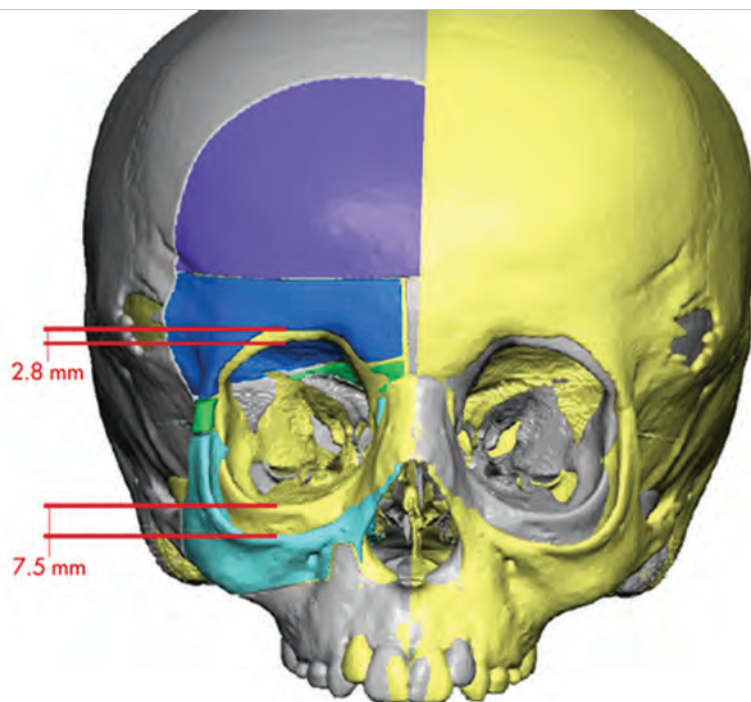


Fig. 5.3 The orbital deformity (green and blue) and the position and shape of a normal orbit (yellow). The exact distance at which the orbit should be elevated and the exact dimension at which the roof needs to be lowered can be measured and the osteotomies planned as depicted.

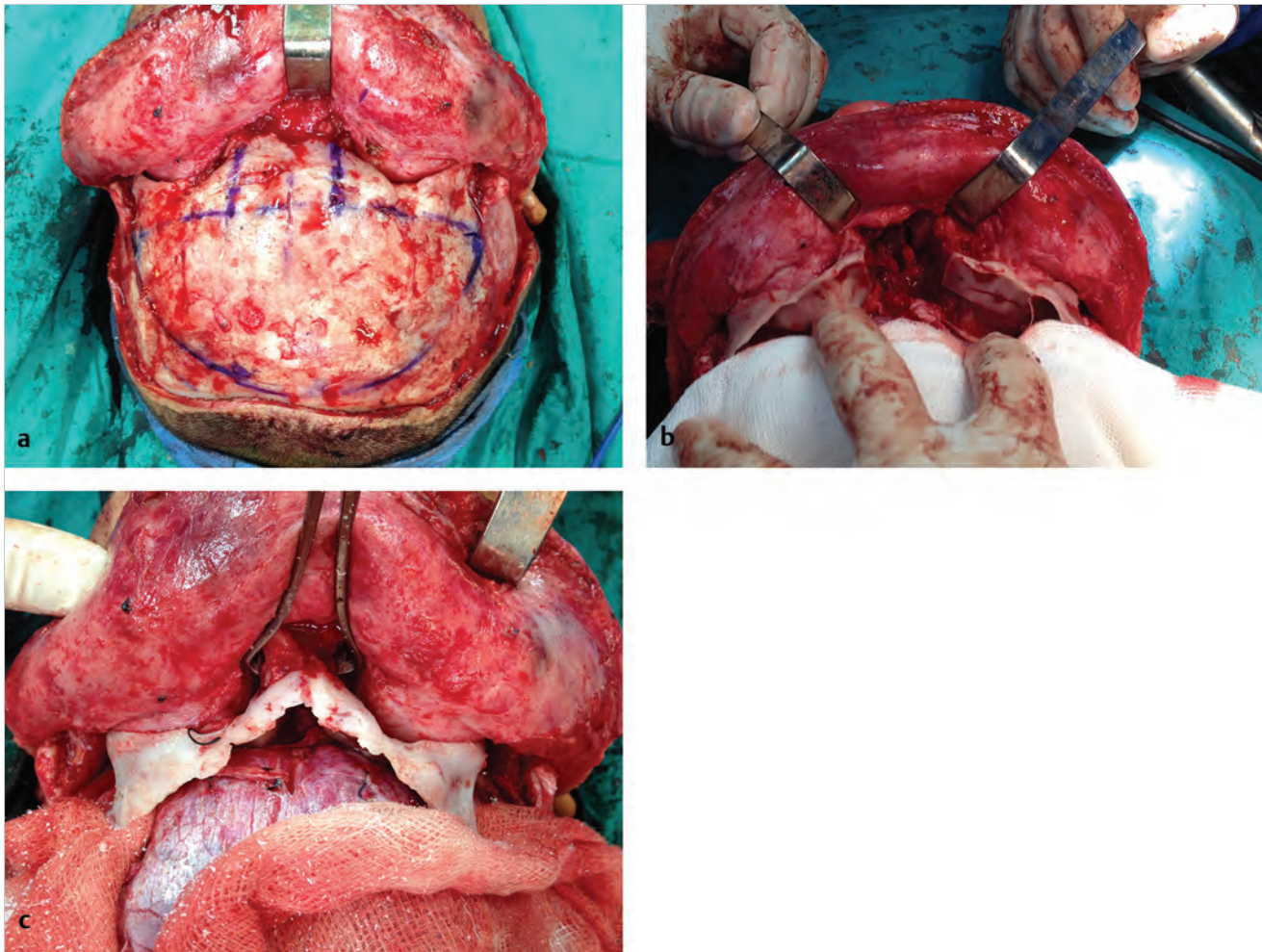


Fig. 5.4 (a) The widened ethmoid bone that will be removed before orbital translocation has been marked in blue ink after coronal incision and exposure. (b) The central segment of bone has been removed after completion of the orbital osteotomies and the bifrontal craniotomy. The nasal ethmoid cells are easily identifiable in the central image between each medial orbital wall. (c) The space between the orbits has been obliterated by mobilization of the entire orbit into close apposition of each other. This bone on bone contact is critical for adequate cranial–nasal separation.

and soft palates in order to cut the perpendicular plate of the ethmoid bone. Full mobilization should allow a side-to-side movement of the monobloc, without much resistance. The previously marked V segment is then resected with a reciprocating saw, ensuring a stable bony attachment for the medial canthi. The reciprocating saw is then used to split the hard palate. A small 2-mm osteotome is placed between the two central incisors to complete the palatal split. At this point, both halves of the bipartition can be brought together at the midline and the intracanthal distance can be measured and verified. Fixation can be achieved with titanium plates or 24-gauge wire. At this point, the distraction device can be placed or the Le Fort I osteotomy can be performed if clinically indicated.

Closure/Soft Tissue Draping

A barrier should be created between the nasal sinus cavities and the epidural space. A pericranial flap is the most useful and first-line tool for proper nasal cranial separation. Fibrin glue can also be placed to fully seal the area. The frontal craniotomy can be fixated in the usual fashion with miniplates. Medial

canthopexies and medial canthal transnasal wiring can be performed and bolstered externally with zeroform gauze, as needed. Closure of the coronal flap will reveal the amount of redundant glabellar skin between the eyebrows. A horizontal mattress suture can be placed within the hair-bearing area of the medial eyebrow, known as the Kawamoto's stitch. The suture is tightened to cause excess soft tissue to bundle in the center of the forehead but position the eyebrows appropriately. Over the following months, the excess tissue will tend to level out and thus avoid a midline forehead scar. Alternatively, the excess skin can be used during a second-stage procedure as a forehead flap for nasal reconstruction seen in rare Tessier 1, 2, or 3 clefts.

5.3 Complications

5.3.1 Intraoperative

Most common intraoperative complications include bleeding, cerebrospinal fluid (CSF) leaks, and dural fistulas. The chances of dural tears and adhesions are often encountered in revisional

or secondary procedures. Meticulous identification of tears or adhesions can prevent most postoperative CSF complications. Preoperative imaging is needed to evaluate any abnormal or large venous sinuses that can potentially be encountered intraoperatively. Visual disturbances can occur postoperatively due to the mobilization of the orbits that results in imbalance of the eye muscles.

5.3.2 Postoperative Management

Patients should be admitted to an appropriate intensive care unit (ICU) postoperatively. Patients are monitored for any signs of increased intracranial and intraorbital pressure. Careful control of intraoperative and postoperative fluid administration will limit the amount of postoperative swelling. Perioperative and postoperative use of steroids may have a role in the management of postoperative swelling as well. We prefer to avoid routine steroid use and use steroids as an adjunct for extubation. We typically start steroid the day before extubation and complete two additional doses after extubation for a total of 24 hours of steroid use. The amount of periorbital and midface well can be quite significant. Without tracheostomy, we prefer 72 hours of postsurgical intubation and sedation. This allows maximal swelling to occur and begin resolution and ensures adequate hemostasis before extubation. Early extubation has led to serious maloutcomes in our experience and thus our abandonment of this practice. When the patient meets clinical parameters for extubation, it should be performed within 5 days of surgery to assist with patient mobilization and enteral feeding. Blindness, diplopia, and anosmia are uncommon complications but should be discussed with the patients. Postoperative imaging is performed to assess the new bony reconstruction as well as any possible complications such as an intracranial hematoma or optic nerve injuries. Postoperative

complications for a facial bipartition are similar to those of the box osteotomies.

5.4 Key Points

- Hypertelorism is a symptom or associated finding that can be accompanied by a multitude of craniofacial conditions that are not genetically or epigenetically related.
- Tessier had classically stressed the fact that hypertelorism is the displacement of the orbital cones in their entirety, manifested by expansion of the central ethmoid sinuses and widening of the sphenoid wing angle to more than 110 degrees.
- The gold standard of correction is a choice between orbital shift using either box osteotomies or facial bipartition.
- Selection of treatment option should be based on the closure pattern and the size of the remaining gap.

Suggested Readings

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6 Rare Craniofacial Clefts

Cassio Raposo, Rizal Lim, and Reza Jarrahy

Summary

The treatment of rare craniofacial clefts starts with making an accurate diagnosis, including an identification of all altered or missing facial soft tissue and skeletal structures.

The ordered Tessier classification system facilitates in this diagnostic process and in surgical planning.

Adherence to basic, established surgical principles of soft tissue and skeletal reconstruction facilitate treatment of even the most complex composite defects.

Surgical intervention is often staged and carried out over many years to achieve a desirable end result.

The best approach to the longitudinal care of patients with rare clefts is through close collaborative multidisciplinary input.

Keywords: rare craniofacial clefts, rare Tessier clefts, facial clefts, encephalocele, facial bipartition, hypertelorism/hypertelorbitism, box osteotomy, cleft lip/cleft lip repair, nasal reconstruction

6.1 Introduction

“In the midst of chaos there is also opportunity” — Sun Tzu. As surgeons, if we look for “common presentations of common diseases” to facilitate our attempts to render accurate diagnoses, develop appropriate treatment plans, and achieve predictable surgical outcomes, then the spectrum of disorders collectively referred to as the rare craniofacial clefts can frustrate even the most skilled and resolute among us. The rare craniofacial clefts represent aberrations of development that challenge our diagnostic skills and our technical abilities. However, like any other complex surgical disease, the key to successful management is making an accurate assessment of the problem, developing a specific set of longitudinal surgical goals, and having an appreciation for how sequential surgical results will evolve over time.

6.2 Classification and Diagnosis

6.2.1 Embryology

Although a comprehensive discussion of human craniofacial embryology is well beyond the scope of this chapter, the complexity of the rare facial clefts and their derivation from failures of embryogenesis merit a brief review of the relevant aspects of in utero facial development.

Much of the critical development of the craniofacial skeleton occurs between the fourth and eighth weeks of gestation, when the endoderm, mesoderm, and ectoderm layers of the face migrate toward the midline. The frontonasal, maxillary, and mandibular processes follow programmed patterns of migration and fusion to create balanced facial features, including the cranial base, forehead, orbits, zygomas, maxilla, nose, stoma, and mandible (► Fig. 6.1). Whether due to a failure of fusion of these migrating processes or due to insufficient migration of mesenchymal cells into the planes of tissue fusion at the

leading margins of these processes—or a combination of both—errors in embryogenesis involving these structures can manifest as vertical, oblique, or transverse clefts.

When Paul Tessier presented his approach to classifying rare craniofacial clefts to the second International Congress on Cleft Palate in Copenhagen in 1973, he offered a comprehensive and concise way to categorize not only the obvious soft tissue deformities but also the underlying aberrant bony anatomy. His logical approach to a set of deformities that seemingly defy logic has since helped us accurately diagnose and appropriately manage these patients.

Dr. Tessier divided the face into various “zones” that are centered on imaginary axes running horizontally through the orbits and vertically through the facial midline. Soft tissue and bony clefts occurring below the orbits are numbered 0 through 8 and those above the orbits are numbered 9 through 14. This numbering system encourages us to evaluate the entire height of the face and thereby more readily identify coincident facial and cranial clefts. When this occurs, the numbers of the individual clefts add up to 14 (e.g., 0–14, 1–13, and 2–12 clefts). Interestingly, Tessier defined clefts of the lower lip, mandible, and associated midline soft tissues of the lower face and neck as number 30 clefts (► Fig. 6.2).

These clefts may be characterized by tissue excess or deficiency; may involve soft tissue, bone, or both; may be unilateral or bilateral; and may be associated with numerous other clefts of diverse orientation. Deconstructing complicated and overlapping cleft patterns according to the diagnostic hallmarks of the individual component clefts, as described later, helps establish accurate diagnoses and develop appropriate treatment plans. In general, physical examination should be complemented by computed tomography (CT) to identify underlying bony abnormalities and by magnetic resonance imaging (MRI) to define the extent of intracranial disease, if appropriate.

Number 0 Cleft

The zero cleft is a midline defect that may involve soft tissue and bone from the central incisors up through the nasal cavity and the perpendicular plate of the ethmoid bone. Mild presentations may be limited to an isolated midline cleft of the lip. With progressive severity, one may note a high and arched or complete cleft palate with absent premaxilla and philtrum, central cleft of the nose with absent columella and nasal septum, hypoplasia of the nasoethmoid complex, arrhinia, and hypotelorbitism. Alternatively, midline tissue excess may result in enlarged and broadened nasal bones, widened nasal septum, and enlarged and laterally or superiorly displaced nasal cartilages. The isolated zero cleft is very rare; it is usually associated with the number 14 Tessier cleft (► Fig. 6.4).

Number 1 Cleft

The number 1 cleft is a vertical paramedian cleft characterized by a cleft lip in the region of the Cupid’s bow and soft tissue

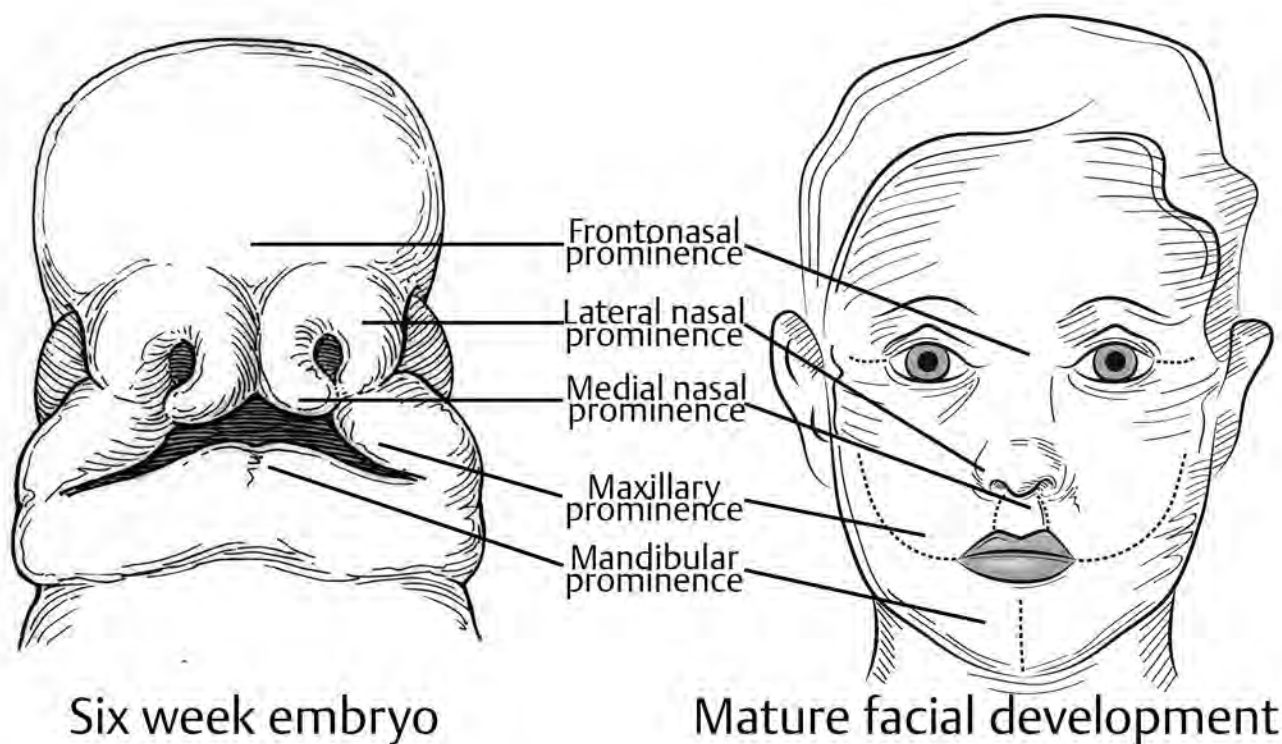


Fig. 6.1 The paired facial processes migrate from lateral to medial during development, contributing to the symmetric features that define the human face.

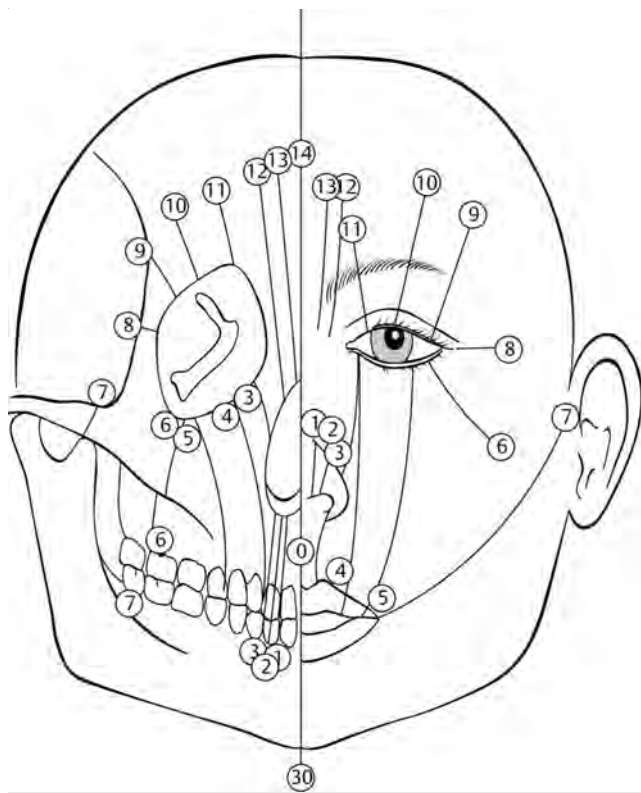


Fig. 6.2 Tessier's classification of rare craniofacial clefts. In his original diagrams, Tessier correlated soft tissue deformities. Right: With their underlying associated bony clefts.



Fig. 6.3 Rare Tessier 30 cleft.



Fig. 6.4 (a) Tessier 0 cleft. This cleft involves the midline caudal to the orbits, resulting in a bifid nasal tip and widened columella but normal interorbital distance and forehead. (b) Tessier 0–14 cleft. The cranial extension of a number 0 cleft is referred to as a number 14 cleft. An encephalocele with associated hypertelorbitism may be present, as shown in this patient. (c) The nasal deformity in this 0–14 cleft is characterized by enlarged and broadened nasal bones, widened nasal septum, and laterally displaced nasal cartilages.



Fig. 6.5 Frontal and lateral photos of a patient with isolated Tessier number 1 cleft. The dome is severely notched, and the upper lateral cartilage on the affected side is absent or other orbital dystopia and the frontal process of the maxilla, resulting in hypertelorbitism or other orbital dystopia.

notching through the dome of the nose that extends toward the medial canthus and medial brow. The nasal defect may include soft tissue fissures over the nasal dorsum or completely missing upper and lower lateral cartilages. Heminasal atrophy or a proboscis may be seen in severe forms. A bony cleft between the central and lateral incisors may extend into the pyriform aperture and cephalad to involve the ethmoid sinuses, the nasal bone, and the frontal process of the maxilla, resulting in hypertelorbitism (► Fig. 6.5).

Number 2 Cleft

In its isolated form, the number 2 cleft is very rare. The cleft lip, such as the number 1 cleft, occurs in the area of the Cupid's bow. The nose is characterized by flattening of the medial third of the nostril, without true notching. The nasal dorsum may be widened, and the septum may be deviated. The palpebral fissure and medial brow are intact but may exhibit inferior displacement or epicanthal folding. The bony cleft begins in the

region of the lateral incisor, and the skeletal fissure extends cephalad into pyriform aperture. The frontal process of maxilla is broad and flat and can be notched. Ethmoid enlargement contributes to hypertelorbitism.

Number 3 Cleft

The number 3 cleft has been described as an oblique cleft, due to the vector of tissue involvement, and as an oro-naso-ocular cleft, due to the facial cavities that it might involve. The soft tissue cleft again begins in the area of the common cleft lip. It continues along the alar base and then obliquely cephalad toward the medial canthus. The cheek tissue deficiency foreshortens the distance from the nasal ala to the medial canthus, causing inferior displacement of this structure. A lower-eyelid coloboma medial to lacrimal punctum may be present, along with agenesis of part or all of nasolacrimal system. Microphthalmia, anophthalmia, and epibulbar dermoids may be seen in this cleft. The cleft in the alveolus occurs between the lateral incisor and the cuspid and extends through the lateral border of the pyriform into the nasal cavity. The bony defect may continue cephalad along the nasomaxillary process to the level of the lacrimal bone and orbital rim and floor, resulting in vertical orbital dystopia. In the most severe cases, absence of bone along this axis results in a confluence of the orbital, maxillary, nasal, and oral cavities (► Fig. 6.6).

Number 4 Cleft

This cleft is also oblique in nature, but unlike the numbers 0 to 3 clefts, the lip defect occurs laterally, between the philtral ridge and the oral commissure. In addition, the defect moves onto the cheek and largely spares the subunits of the nose, despite the shortened alar-ocular distance. The medial canthus is largely spared, as the cleft transitions onto the lower eyelid, lateral to the punctum. The lacrimal drainage structures are therefore intact but often dysfunctional. The globe can show anophthalmia, microphthalmia, or normal anatomy. The alveolar defect also occurs between the lateral incisor and cuspid, but the pyriform is spared. The bony cleft extends through the maxilla

medial to the infraorbital foramen and then into the orbit. Herniation of orbital contents yields a hypoplastic and dystopic orbit. In the complete form of the number 4 cleft, the oral, maxillary, orbital cavities are confluent, for which reason this cleft has been referred to as the oro-ocular oblique cleft (► Fig. 6.7).

Number 5 Cleft

The number 5 cleft is the rarest of the oblique facial clefts and is rarely seen in isolation. Its clinical characteristics are similar to those of the number 4 cleft. The maxillary defect occurs *lateral* to the infraorbital foramen and then across the rim and onto the floor of the lateral orbit.

Number 6 Cleft

The number 6, 7, and 8 rare craniofacial clefts often coincide as Treacher Collins syndrome (TCS), and the isolated number 7 cleft underlies the findings associated with craniofacial microsomia. Both of these clinical entities are described in detail elsewhere in this text and will therefore not be discussed in depth here. However, there are relevant aspects of these individual clefts that merit independent discussion.

In contrast to the syndromic presentation, which is characterized by absence of the zygoma and its arch, the isolated



Fig. 6.6 Tessier number 3 cleft. The cleft lip is located in the region of the Cupid's bow. The alar base is involved, and the cleft runs toward the medial canthus, which is inferiorly displaced. Note the associated anophthalmia in this case.



Fig. 6.7 Bilateral Tessier number 4 cleft. The nose is largely spared, as is the medial canthus on the patient's right, in this bilateral asymmetric case. Note the left-sided medial canthal distortion, anophthalmia, and orbital dystopia.

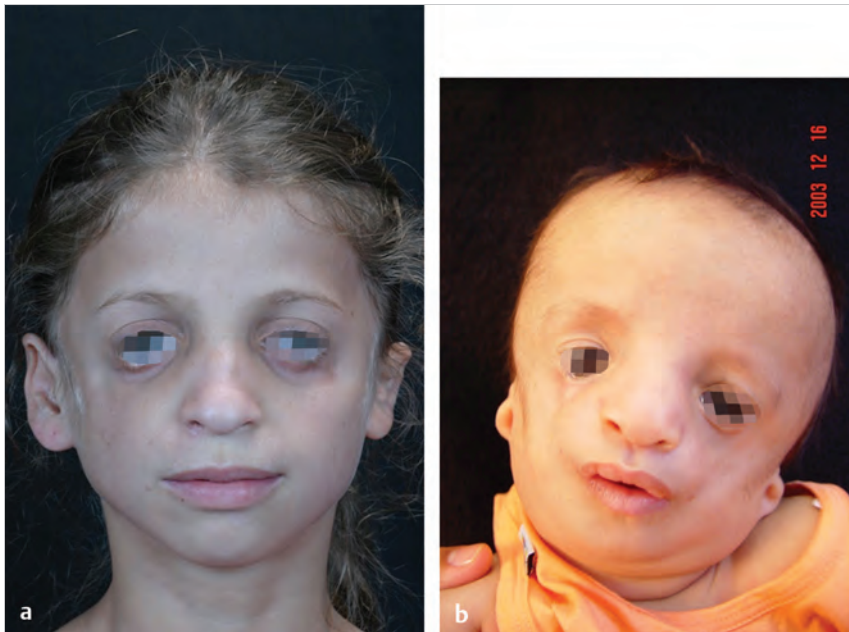


Fig. 6.8 Isolated Tessier number 6 cleft. (a) This cleft is considered by some to represent an incomplete form of Treacher Collins syndrome. Lower-eyelid colobomas are seen, but the lids and lateral canthi are less severely dystopic compared with the syndromic presentation (b) due to greater underlying skeletal support.

number 6 cleft is defined by a present but hypoplastic zygomatic body and an intact zygomatic arch. Soft tissue manifestations are similar to those seen in patients with TCS but are milder in nature (► Fig. 6.8).

Number 7 Cleft

In addition to its role in TCS, the number 7 cleft is the most common isolated rare craniofacial cleft. The numerous ways in which it has been historically referred to indicate the variability in its clinical manifestation: hemifacial microsomia, craniofacial microsomia, microtia, otomandibular dysostosis, first and second branchial arch syndrome, and oromandibuloauricular syndrome. Macrostomia is the hallmark feature of the cleft, but soft tissue involvement can be diffuse, ranging from small preauricular skin tags to significant deficiency and distortion of the auricle, cheek, tongue, soft palate, parotid gland, and facial musculature. The cardinal bony defects are centered on the zygomaticotemporal suture, with resultant hypoplasia or absence of the zygoma or mandible (► Fig. 6.9).

Number 8 Cleft

An isolated number 8 cleft is a rare entity. Centered on the zygomaticofrontal suture, the cleft begins at the lateral palpebral fissure and extends to the temporal region. Soft tissue furrows, discontinuity of the orbicularis oculi ring, colobomas, and dermatoceles occur in the region of the lateral canthal tendon insertion. The bony defect manifests as bone loss of the lateral orbital rim at the level of the suture.

Number 9 Cleft

An isolated number 9 cleft is extremely rare. The cleft is located at the superolateral angle of the orbit. The upper eyelid may exhibit full-thickness tissue loss between its median and lateral thirds. This defect may extend through the brow and obliquely



Fig. 6.9 Bilateral Tessier number 7 cleft. Macrostomia is the hallmark of the isolated number 7 cleft. This patient demonstrates asymmetric soft tissue and mandibular involvement.

toward the hairline and may involve the underlying orbit and the forehead (► Fig. 6.10).

Number 10 Cleft

This cleft represents the cranial extension of the number 4 cleft and may manifest as a coloboma of median third of the eyelid,



Fig. 6.10 Tessier number 9 cleft.

with continuation to the hairline. A whorl of hair-bearing scalp may be seen in the frontal region in the region of the cleft. The underlying skeletal defect can include absence of the median orbital rim, roof, and adjacent frontal bone, with resultant fronto-orbital encephalocele. In this setting, the orbit is laterally and inferiorly rotated (► Fig. 6.11).

Number 11 Cleft

The number 11 cleft typically occurs with the number 3 facial cleft. It passes through the medial third of the upper eyelid and eyebrow, extending to the frontal hairline. It can involve the orbit, causing a defect of the medial orbital rim, or the ethmoid complex, resulting in orbital hypertelorbitism (► Fig. 6.12).

Number 12 Cleft

The number 12 cleft represents the cranial extension of the number 2 cleft. It passes either through the frontal process of the maxilla or between this process and the nasal bone. Because of the increased width of the ethmoid complex,

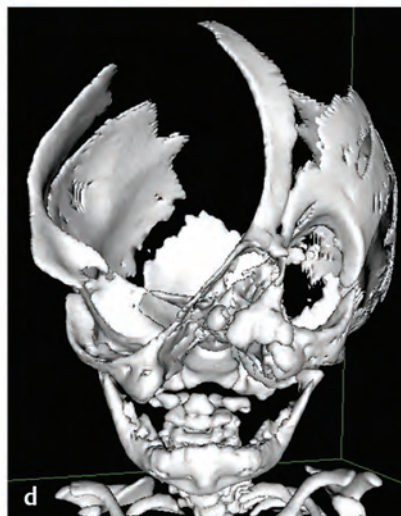


Fig. 6.11 (a) Isolated Tessier number 10 cleft with associated encephalocele. The upper eyelid and brow are distorted, and the hypertelorbitism is characterized by lateral and inferior displacement of the orbit. (b) By comparison, this number 10 cleft has an associated incomplete form of a number 4 cleft, with a previously repaired cleft lip, mild perinasal deformity, shortened naso-ocular distance, and inferiorly displaced medial canthus. (c) The number 10 cleft can be associated with massive encephaloceles, as in this case with a complex pattern of multiple severe coincident bilateral clefts. (d) CT scan shows the large fronto-orbital defect.

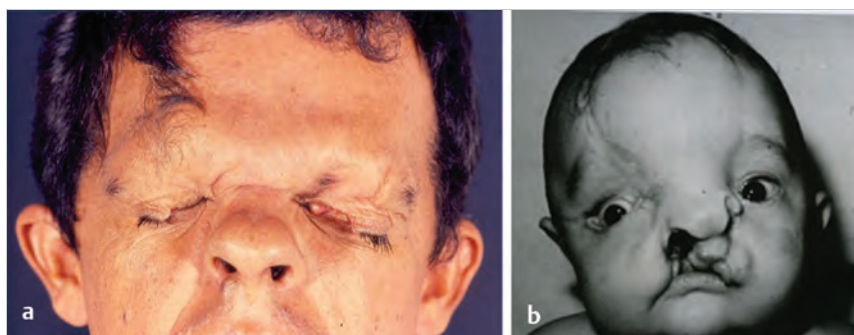


Fig. 6.12 (a) A rare bilateral Tessier number 11 cleft involves the medial portions of the upper eyelid and brows, extending toward the frontal hairline. (b) More commonly, the number 11 cleft occurs with the number 3 cleft as a Tessier 3-11 cleft. In this asymmetric bilateral case, involvement of the lip, nose, medial canthus, orbit, brow, and forehead is more severe on the patient's right side.



Fig. 6.13 (a) A Tessier 1-13 cleft with mild nasal deformity but significant hypertelorbitism with associated encephalocele. (b) In this case, the 1-13 cleft is coincident with an ipsilateral number 10 cleft, with associated anophthalmia and orbital deformity.

hypertelorbitism is a hallmark characteristic. The olfactory organs and cribriform plate are typically structurally normal. The overlying soft tissue may include brow irregularities immediately lateral to its medial edge.

Number 13 Cleft

Displacement of an otherwise-uninterrupted medial brow is characteristic of a number 13 cleft. The underlying skeletal anomaly is consistent with cranial extension of the number 1 cleft, passing between the nasal bone and the frontal process of the maxilla through the ethmoids and cribriform plate, which may be inferiorly displaced in the setting of a concurrent encephalocele. Collectively, these skeletal components can result in severe hypertelorbitism (► Fig. 6.13).

Number 14 Cleft

The zero cleft continues onto the central forehead as the number 14 cleft, which has a variety of presentations. If the cleft is characterized by tissue agenesis, clinical manifestations can include hypotelorbitism, cyclopia, ethmocephaly, or cebocephaly. Associated holoprosencephaly heralds a poor overall prognosis and limited life expectancy. When, by comparison, the cleft is characterized by incomplete migration of the frontonasal process, a resulting bifid cranium or frontal encephalocele may maintain

the orbits in a lateralized position, with resultant hypertelorbitism. In this setting, an enlarged or duplicated crista galli, widened olfactory grooves, inferiorly displaced cribriform plates, and enlarged ethmoids are also characteristic (► Fig. 6.4).

Number 30 Cleft

Finally, the number 30 cleft encompasses clefts of the lower lip and mandible that fall on the same axis as the number 0 cleft. These may involve the tongue, hyoid bone, and midline structures of the neck to the level of the sternum.

6.3 Nonoperative Management

Definitive management of rare craniofacial clefts is surgical in nature. Nonsurgical care of patients with rare craniofacial clefts is generally supportive in nature and helps facilitate successful surgical outcomes. Under ideal circumstances, patients are treated in a multidisciplinary craniofacial clinic, with close coordination among all involved caregivers.

6.3.1 Feeding and Nutrition

In the immediate postnatal period, the rare facial clefts that involve the upper lip and/or palate may pose challenges to

feeding and potentially contribute to dehydration and malnutrition. Parents must be educated and guided on techniques and materials that can facilitate adequate oral feeds. Consultation with an occupational therapist who is experienced in the management of children with facial clefts may be indicated. This support should coincide with a larger more holistic approach to providing longitudinal psychosocial support to patients and their families.

6.3.2 Neurologic Assessment

Children with rare craniofacial clefts with any suggestion of encephalocele, holoprosencephaly, or other structural cranial deformity should undergo neurologic and neurosurgical assessments. These evaluations should include radiographic examination of the craniofacial skeleton via CT scan and of the brain via MRI.

6.3.3 Dentistry/Orthodontia

Virtually all patients with rare craniofacial clefts will require attention from a pediatric dentist and an orthodontist during their growth and development. Orthodontic treatment should be tailored to each individual's needs, as hypodontia, alveolar misalignment, malocclusion, and even orthodontic sequelae of surgical interventions can have aesthetic and functional consequences on occlusal relationships.

6.3.4 Speech

Cleft patients may have difficulties with speech, which stems from structural abnormalities or from learned misarticulations. The speech pathologist plays a critical and ongoing role in identifying undesirable speech patterns early in language development, providing speech therapy, and monitoring speech patterns throughout childhood and young adulthood.

6.4 Operative Management

Because of the wide breadth of clinical diversity that is inherent to rare craniofacial clefts, it is impossible to define specific algorithms or protocols for any one specific cleft entity. However, Tessier's ordered classification system allows us to draw upon well-established basic principles of cleft, craniofacial, and general plastic surgery to address the complex defects that we encounter in these patients. Described later is, first, a summary of general approaches to surgical management, followed by some cleft-specific considerations. Although not comprehensive, this overview provides a template for developing surgical treatment plans.

6.4.1 Soft Tissue Reconstruction

Perioral Reconstruction

Perioral reconstruction is fundamental to all of the facial clefts that involve the soft tissues of the upper lip (and, in the case of the number 30 cleft, the lower lip). Repair follows the principles of common cleft lip repair that are discussed at length elsewhere in this textbook, including the use of presurgical

nasoalveolar molding devices. The goals of surgery are to establish continuity of the three tissue layers of the lip—skin, mucosa, and muscle—with reconstitution of the muscular sphincter, creation of the philtral unit, shaping of the nose, and aesthetic placement of scars. Cleft palate repair in patients with rare facial cleft should follow the common principles of three-layer intravelar veloplasty and two-layer closure of the hard palate.

Periorbital Reconstruction

The upper and lower eyelids and medial and lateral canthal tendons are involved to varying degrees in rare cleft deformities and are particularly vulnerable to deformation in the lateral facial oblique clefts. Eyelid coloboma repair follows conventional principles. Complete colobomas with associated risk for corneal exposure and keratopathy must be approached with greater urgency than partial defects with more limited functional deficit. Caudal displacement of the medial and lateral canthi calls for a variety of corrective measures that include canthopexy and canthoplasty, Z-plasty, and other interdigitating local tissue transpositions. Microphthalmia should be managed in coordinated fashion by a pediatric ophthalmologist.

Nasal Reconstruction

The treatment of rare facial clefts demands a thorough understanding of the concept of the aesthetic units of the face, described by Gonzalez-Ulloa. Burget and Menick applied this concept to develop a comprehensive approach to nasal reconstruction, including the need for trilaminar nasal reconstruction, the use of local and regional tissue rearrangement for lining and coverage of defects, and the placement of scars between distinct anatomical areas to optimize scar aesthetics. Microsurgical techniques may also be indicated in complex defects where the supply of local tissue is scarce.

6.4.2 Bone Reconstruction

In addition to the standard techniques that one would use to consolidate the maxillary alveolus or reconstruct orbital defects with autologous bone graft, certain aspects of complex skeletal reconstruction deserve specific attention, as they relate to the management of rare craniofacial clefts.

Hypertelorbitism Repair

Hypertelorbitism is a potential trait of any of the rare clefts associated with skeletal defects above the horizontal axis of the orbit. Clefts 9 through 14 can present with either symmetric or asymmetric hypertelorbitism, in which case the major goal of surgical correction is medialization of the orbit(s). The commonly practiced techniques of hypertelorbitism repair are well described in the literature, as are varying opinions on the ideal age to perform the surgery to avoid long-term relapse of the repositioned orbit. In general, older patients with adequate occlusion are good candidates for an orbital box osteotomy procedure. By comparison, skeletally immature patients and patients with inverted-V maxillary morphology benefit from a facial bipartition procedure. Other considerations that impact the choice of technique include the amount of cribriform prolapse into the ethmoid complex and the degree of

hypertelorbitism, as originally classified by Günther and Tessier. For patients with an intact calvarium and skull base, hypertelorbitism may be treated by medialization of the orbits via a subcranial Le Fort III osteotomy.

Box Osteotomy

In this combined intracranial/extracranial approach, coronal and gingivobuccal sulcus incisions are used to obtain subperiosteal exposure of the frontal bones, orbits, and midface. A bifrontal craniotomy is performed. The interdacryon distance is measured with calipers. Circumferential orbital osteotomies are made, with preservation of the medial canthal insertions. The zygomaticomaxillary and nasomaxillary buttresses are also cut. Following adequate intracranial midline dissection and retraction, a central frontoethmoidal segment is resected. The width of this resection is determined by the planned amount of correction. The orbits are translocated medially, and the vertical buttresses and nasofrontal process are rigidly fixed with titanium plates and screws. A pericranial flap is raised and sutured down to the cranial base in the midline before closure (► Fig. 6.14).

Facial Bipartition

In contrast to the box osteotomy technique, the zygomaticomaxillary and nasomaxillary buttresses are left intact in a facial bipartition. Pterygomaxillary, septal, and median palatal osteotomies are added to the bone cuts, described earlier, to allow complete midface mobilization. Rowe disimpaction forceps are used to down fracture the midface following completion of the osteotomies. A wedge of central nasal, frontal, and ethmoid bone is removed, and the hemifacial segments are rotated toward the midline. Rigid fixation is applied, and autologous bone grafts are used to stabilize the advanced lateral orbital rims and zygomatic arches. When indicated, a cantilever nasal bone graft is placed and rigidly secured (► Fig. 6.15).

Treatment of Encephalocele

When the cleft is associated with an encephalocele, a feature that is commonly found in the number 10 cleft but is also seen in number 9 and 11 through 14 clefts, neurosurgical intervention must be incorporated into the longitudinal surgical plan. The encephalocele can be treated by excision of abnormal, nonfunctioning brain tissue and simultaneous restoration of calvarial integrity with autologous bone grafts. Secondary hypertelorbitism correction, as described earlier, is planned for a later stage (► Fig. 6.16).

The general principles discussed earlier can be applied to any applicable defect that falls on the spectrum of rare craniofacial clefting. An elaboration of some specific considerations that apply to individual clefts is discussed as follows.

Median and Paramedian Clefts: 0–14, 1–13, and 2–12 Clefts

Surgical treatment depends on the extent of the deformity. Treatment of the facial and cranial components should be staged, starting with reconstruction of the cleft lip deformity at

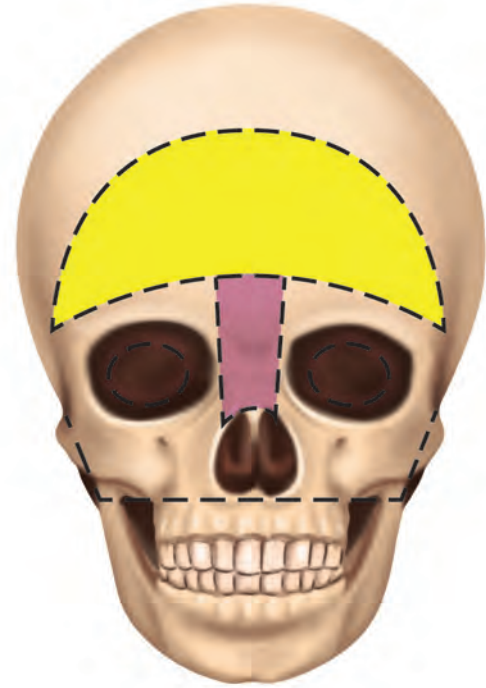


Fig. 6.14 Box osteotomy.

approximately 3 months of age according to standard cleft lip repair principles. Subsequent repair of the nose can be accomplished by a combination of local and regional tissue advancement, rotation, and transposition flaps, in combination with composite or cartilaginous auricular grafts for support. More complex reconstructions are necessary in clefts with cranial extension that affect the orbits and forehead, following the guidelines for management of hypertelorbitism with or without encephalocele, as described earlier. When there is significant deformity of the nose, requiring an extensive amount of soft tissue for coverage, initial correction of the hypertelorbitism may provide additional mobile tissue to recruit for nasal reconstruction.

Numbers 3–11 Cleft

The overall goals of care are to provide protective soft tissue coverage to the globe; restore function to the nasolacrimal structures; correct the position of the involved medial canthi; establish acceptable nasal form and function; increase the foreshortened naso-ocular distance; and restore bony separation between the oral, nasal, and orbital cavities. The initial care of the patient should focus on ocular protection and minimization of infection from a blind-ending nasolacrimal system. Cleft lip and nose repair, reconstruction of eyelid colobomas, and closure of the soft tissue portions of the naso-orbitomaxillary cleft can begin at 3 months of age. The deficient naso-ocular distance can be addressed with multiple interdigitating Z-plasties, regional tissue transfer, or undermining and advancement of

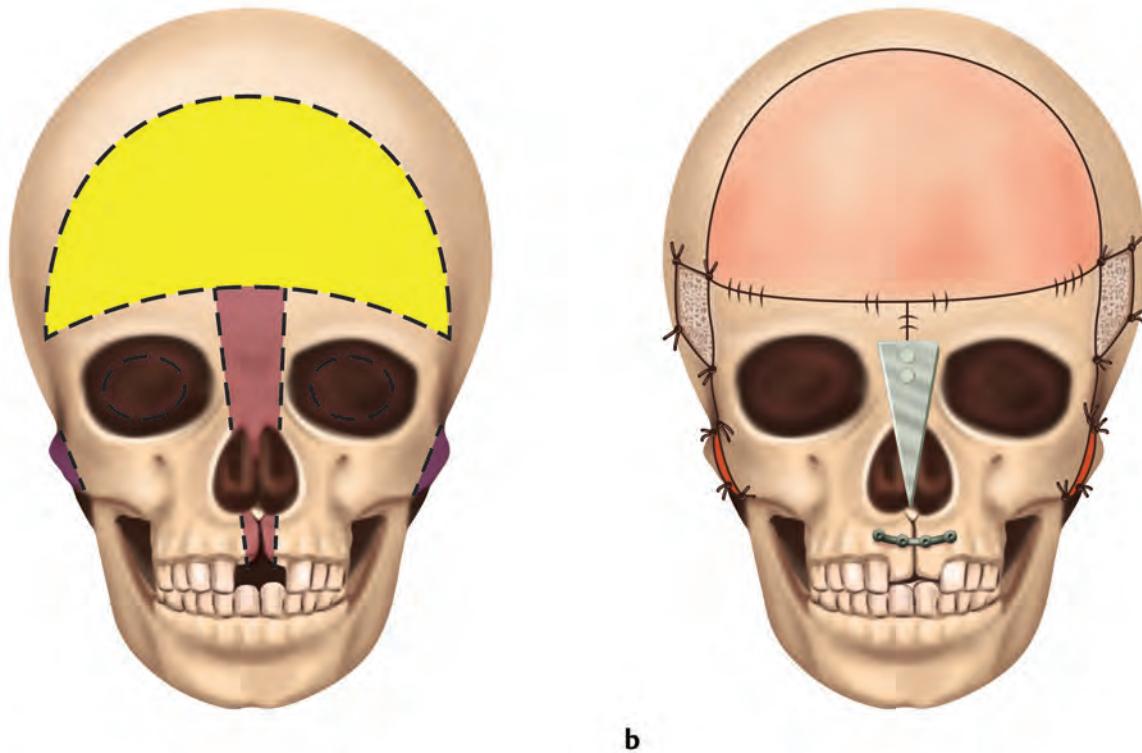


Fig. 6.15 Facial bipartition.

the superficial musculoaponeurotic system of the face as a Mustarde's flap. Lower eyelid and medial canthal reconstruction, also using adjacent or local tissue transfer, can be performed concurrently or deferred until a later stage.

Bony reconstruction typically follows soft tissue correction. This may include bone grafting for orbitomaxillary defects, hypertelorbitism correction, or both. Sources of autologous bone graft include iliac crest, split calvarium, and rib. Bone graft resorption is well described, regardless of the graft source, and may necessitate secondary bone grafting.

Numbers 4–10 Cleft

The principles governing soft tissue reconstruction of the 4–10 cleft are very similar to those described for the 3–11 cleft, including goals, techniques, and timing. Ocular protection is a critical early priority. One significant difference in the cleft lip repair technique of the number 4 cleft is to discard the normal lip tissue that falls between the cleft laterally and the ipsilateral philtral column medially. Although it is seemingly counterintuitive to discard normal tissue in the setting of a disease characterized by severe tissue deficit, this maneuver is necessary to place the scar within the appropriate facial subunit. Cheek advancement, lid switch, nasolabial flap, and other regional flaps, all facilitate cheek, lower eyelid, and medial canthus reconstruction.

Orbital reconstruction in cases of anophthalmia is a staged process. The first stage involves the creation of a palpebral

fornix by lining the inner surface of the chosen eyelid's reconstructive flap with a buccal or septal mucosal graft. The volume of the hypoplastic orbit must then be progressively increased in anticipation of an ocular prosthetic. This can be accomplished with implanted tissue expanders or with serial placement of orbital conformers of increasing size.

Management of encephaloceles, which can be particularly large in number 10 clefts, follows the previously described principles. Repair typically requires a combined intracranial/extracranial approach and adheres to the standard paradigm of three-layer reconstruction of dura, bone, and skin. A facial bipartition can be performed at the same time to derotate the orbit. Correction of the encephalocele and hypertelorbitism usually provides redundant soft tissue that can be used in secondary repairs or revisions of the original cheek, eyelid, canthus, or nasal reconstructions.

Numbers 5–9 Cleft

Similar to the other oblique facial clefts, neonatal corneal exposure should be carefully managed to avoid permanent ocular injury. As noted earlier, soft tissue considerations take early precedence. **W-** or **Z-**plasty of the cheek is a useful approach to reconstruction of the soft tissue furrow caudal to the eyelid. The lower-eyelid coloboma can be repaired with local tissue rearrangement such as the Fricke's flap, upper eyelid switch flap, or advancement of the uninvolved portions of the lower lid. Abnormalities of the lateral canthal tendon that are



Fig. 6.16 (a) Transillumination of a patient with the complex number 10 cleft, shown previously in Figure 10, shows the magnitude of the encephalocele defect and the tenuous coverage. (b,c) Combined intracranial/extracranial approach allows repair of the encephalocele and dura, assessment of the calvarial defect, and repair. In this case, autologous cranial bone and resorbable mesh plates were used to reconstruct the forehead and provide protective coverage to the brain. (d) Immediate and (e) short-term follow-up before secondary surgical corrections.

associated with the cleft should be treated with lateral canthopexy at the time of lower-lid repair. All the principles detailed earlier regarding management of anophthalmia, microphthalmia, orbital defects, hypertelorbitism, and encephalocele in 4–10 and 3–11 clefts all apply to 5–9 clefts.

6.4.3 Lateral Clefts

Number 6 Cleft

Correction of the cheek soft tissue furrow and eyelid coloboma that overly the deficient orbitomalar skeleton can be obtained early in childhood with a combination of Z-plasties and upper-

eyelid transposition flaps. Bone grating to the zygoma, zygomatic arch, and orbit can be performed later in childhood when adequate sources of autologous bone are available. Secondary soft tissue augmentation is accomplished using standard autologous structural fat-grafting techniques.

Number 7 Cleft

Initial macrostomia repair should not only match the transverse length and wet/dry vermilion orientation of the lip on the uninvolvement side but also reorient the zygomaticus major, risorius, and depressor anguli oris muscles to create the absent modiolus. Both straight-line and Z-plasty skin closures are acceptable.

Microtia repair by using autologous rib cartilage, porous polyethylene implants, or osseointegrated prostheses are all well described.

The technique and timing of skeletal reconstruction of the deficient mandible depend on the degree of hypoplasia, mandibular growth, and secondary effects on the maxilla. Options include rib grafting for the congenitally absent mandible, distraction osteogenesis for the significantly shortened mandibular body or ramus, and conventional orthognathic correction for skeletally mature patients with mandibular asymmetry and malocclusion. Free vascularized bone grafts have also been described for mandibular agenesis.

Number 8 Cleft

Soft tissue reconstruction involves excision of the cleft tissue and repair with tarsopalpebral flaps and laterally based cutaneous flaps. A lateral canthoplasty is performed and covered with adjacent soft tissue.

6.5 Complications

In any complex disease pattern requiring multiple intricate surgical interventions, adverse events are a reality; these may be related either to bone or to soft tissue reconstruction. Encephalocele and hypertelorbitism repair are associated with numerous complications that are inherent to combined intracranial and extracranial procedures. These include death, stroke, hemorrhage, cerebrospinal fluid leak, meningitis, vascular injury, and osteomyelitis. Orbital translocation carries the additional risks of optic nerve injury and inadvertent medial canthal avulsion. Frontoethmoidal dissection and resection carry the risk of olfactory sensory loss. Closely coordinated multidisciplinary perioperative planning and meticulous intraoperative technique mitigate these risks.

Soft tissue complications may occur more frequently but are less severe in nature. Partial or total flap necrosis, soft tissue infections, cicatricial deformities, and recurrent asymmetries may all contribute to the need for surgical revision.

6.6 Conclusion

The craniofacial skeleton is a three-dimensional anatomical structure that is constantly evolving within the fourth dimension of time. In patients with rare craniofacial clefts, specific anatomical elements of the involved soft tissues or skeleton may be absent, malformed, or misplaced. Appreciation of the embryologic theories underlying pathogenesis helps us appreciate the severity of the deformities that we encounter. The Tessier classification system allows us to evaluate patients systematically and logically. A multidisciplinary and systematic approach can yield excellent results. However, the ever-changing nature of facial tissues renders construction of missing elements or reconstruction of aberrant structures much more difficult: if the surgeon's goal is to establish long-lasting balance and harmony in a face that is devoid of either, we must predict not only how our results will appear 1 month or 1 year after surgery but also how those results will evolve over subsequent

decades. Although we have some control over our surgical interventions, we have little control over how our patients heal and how scarring contributes to undesirable secondary effects on facial growth and function. A forward-thinking perspective and consistent longitudinal care improve our chances of obtaining long-term success.

The care of patients with rare craniofacial clefts is challenging and often humbling. When faced with seemingly impossible reconstructive challenges, we must rely on fundamental surgical principles, collaboration, and persistence to provide the level of care that these children require and deserve. Although we consistently strive to improve our surgical results with new and innovative techniques, we must not forget that a far more important measure of success than improvement in the patient's physical appearance is whether our involvement in their care has served to, in the words of Tagliacozzi, "buoy up the spirit, and help the mind of the afflicted."

6.7 Key Points

- The treatment of rare craniofacial clefts starts with making an accurate diagnosis, including an identification of all altered or missing facial soft tissue and skeletal structures.
- The ordered Tessier classification system facilitates in this diagnostic process and in surgical planning.
- Adherence to basic, established surgical principles of soft tissue and skeletal reconstruction facilitates the treatment of even the most complex composite defects.
- Surgical intervention is often staged and carried out over many years to achieve a desirable end result.
- The best approach to the longitudinal care of patients with rare clefts is through close collaborative multidisciplinary input.

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7 Facial Fractures

S. Alex Rottgers and Joseph Edward Losee

Summary

Treatment of facial fractures in pediatric patients differs from adults. Different mechanisms of injury, dental anatomy, facial proportions, and bone quality result in unique injury patterns not seen in older patients. Treatment plans must account for the possibility that the injury and/or operative treatment can alter future facial growth. Additionally, with future growth and dental eruption, the pediatric facial skeleton is adaptive and minor malocclusions and subtly displaced fractures may improve with time. Subsequently, pediatric facial fractures are treated less frequently with open reduction and internal fixation, and are more frequently treated conservatively. Open treatment is sometimes required. In these instances, careful considerations are needed to protect immature dental structures and to avoid or remove permanent implants that may restrict growth. Basic principles of patient assessment and operative/non-operative management of pediatric craniofacial trauma are discussed with special attention brought to the patterns of growth seen in the facial skeleton and how the anatomy at different stages of development should affect a surgeon's clinical decision making when managing facial trauma in young patients. Unique fracture patterns, fixations techniques, and potential complications seen only in the pediatric population are discussed.

Keywords: pediatric, facial fractures, craniofacial trauma, orbital fracture, nasal fracture, naso-orbital-ethmoid fracture, Le Fort fracture, zygomaticomaxillary complex fracture, mandible fracture, facial growth, facial development

7.1 Introduction

Management of facial fractures in pediatric patients differs from that of their adult counterparts. Variations in anatomy, different daily activities, and the presence of ongoing growth account for these differences. Facial trauma is more common in children than in adults, but the facial skeleton is fractured less frequently. Pediatric patients account for 15% of facial fractures. When fractures occur, they affect areas of the face with different frequencies, occur in different patterns, and are more commonly greenstick fractures. The treatment goal is not simply to reestablish normal anatomy and function but to do so without disrupting future growth. The plasticity of the growing skull allows significant remodeling with growth, so more conservative modalities are often successful. When open reduction and fixation is necessary, surgeons must avoid injury to developing dental structures and growth sites, consider the effect that wide subperiosteal undermining may have on future growth, and recognize that fixation hardware has the potential to restrict growth or migrate into the bone or cranium. The specter of the fourth dimension haunts every surgeon caring for these injuries and must weigh on every management decision.

7.2 Patterns of Craniofacial Growth

Understanding facial development is key to understanding fracture patterns in pediatric patients and anticipating how they might affect future growth. At birth, the cranium is significantly larger than the face, with a proportion of 8:1. With pneumatization of the paranasal sinuses and eruption of the dentition, this proportion changes over time. At 5 years of age, this ratio shifts to 4:1 and reaches a ratio of roughly 2:1 at skeletal maturity. The majority of cranial growth is completed by the age of 3 years. The brain's volume triples in the first year of life. The neurocranium is 25% of its adult volume at birth, 75% at 2 years of age, and 95% at 10 years of age. Growth is much more delayed in the facial skeleton, with the face achieving only 65% of its adult proportions by 10 years of age.

The cranium and the upper face grow as a result of brain and ocular expansion. During the early rapid expansion of the cranium, growth is dependent on bone deposition at the cranial sutures. As this process slows, further growth results more from appositional bone deposition. Although some growth remains, expansion of the cranium and orbits are nearly completed by 6 years of age.

The maxilla and midface continue to grow until roughly 12 years of age. Flattening of the cranial base with early brain development, growth at cranial base synchondroses, transduction of forces from nasal septal growth via the septovomerian ligament, and vertical alveolar elongation with dental eruption, all account for the forward and downward displacement of the maxilla. In addition, pneumatization of the paranasal sinuses contributes to maturation of the midface. All of the primordial paranasal sinuses have formed at the time of birth, but only the maxillary sinus is radiographically visible and clinically relevant. Ethmoid air cells begin to pneumatize in the first 2 years of life and continue to enlarge thereafter. The sphenoid and frontal sinuses are not radiographically apparent until after the age of 6 years and will continue to enlarge in early adulthood.

The mandible begins as two separate bones joined by a cartilaginous suture at the symphysis, which fuses during the first year of life. Mandibular growth continues with the rest of the face during childhood. The mandible continues to grow until skeletal maturity. Growth occurs both at the condylar head, which functions as a growth site, and through centripetal expansion. Condylar growth contributes to vertical and anteroposterior (AP) enlargement of the mandible. Eruption of dentition also adds to the vertical height of the alveolar segments, as it does in the maxilla.

7.3 Varied Fracture Characteristics in Children versus Adults

The high craniofacial proportion in early life and the lack of pneumatized sinuses account for the different fracture patterns seen in younger patients. In children younger than 6 years, the

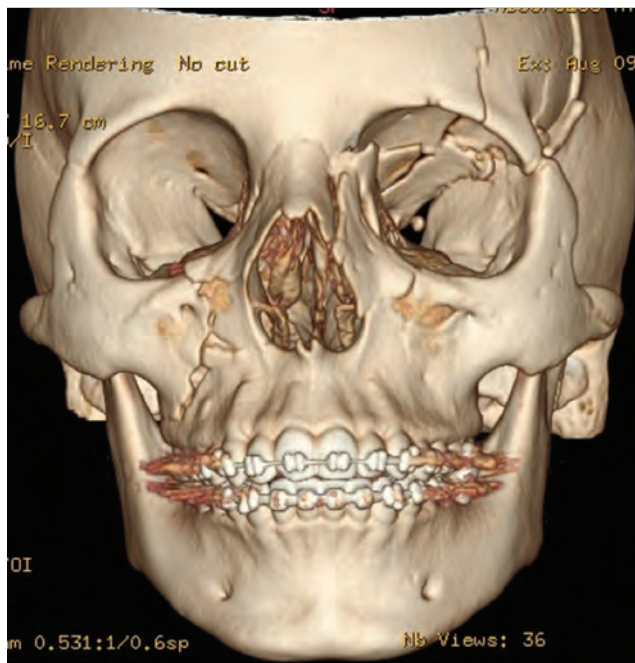


Fig. 7.1 Oblique craniofacial fracture. Midface fractures with Le Fort pattern are uncommon when patients are in primary and early mixed dentition. Young children lack pneumatization of the perinasal sinuses and have proportionally larger orbits and crania. Cranial fractures are more common in these patients. Injuries may propagate through the orbital roof and into the buttresses surrounding the pyriform aperture and malar prominence. These fractures typically follow an oblique trajectory, such as that seen in this patient with a left frontal bone fracture that extended into the orbital roof, ipsilateral nasomaxillary buttress, and the contralateral inferior orbital rim and zygomaticomaxillary buttress.

most common fractures are orbital and cranial fractures. These structures are proportionally larger and have greater prominence in the immature facial skeleton. Without a frontal sinus to absorb the energy of impact, fractures are prone to propagation into the frontal bone and along the orbital roof toward the apex. Fractures may also extend in an oblique trajectory into the midface, crossing either the nasal cavity or the buttresses surrounding the zygoma (► Fig. 7.1).

The midface is shielded from trauma and is more elastic and resistant to fracture in young children. Younger patients rarely exhibit the classic Le Fort pattern fractures seen in adults and adolescents. Immature bone is more cancellous and is less mineralized. The midface of younger patients is void of sinuses and is filled with dental follicles. Without aerated maxillary sinuses, force on the maxilla is transferred to the alveolus and results in dentoalveolar trauma. The palatal suture remains patent into adolescence and results in a higher incidence of palatal split fractures in children.

The lower mineral content and greater percentage of cancellous bone often result in deformations and greenstick fractures in children. When fractures require open reduction, they may require osteotomy to allow mobilization and proper reduction. In addition, dental eruption and remodeling can often overcome minor deformities and occlusal irregularities. Fractures that would clearly require open treatment in adults can be

successfully managed with conservative measures and subsequent orthodontic correction in children. A new balance must be found while treating pediatric facial fractures when deciding what constitutes an operative case.

7.3.1 Diagnosis

Initial Assessment and Physical Examination

Evaluation of pediatric patients with facial trauma should begin with a comprehensive evaluation according to the principles of Advanced Trauma Life Support (ATLS) to avoid missing concurrent significant or life-threatening injuries. Another serious injury is present in 56% of patients. The most common associated injuries are neurologic, which occur in 47% of patients. Half of these injuries are concussions. Patients must be adequately screened and referred for appropriate follow-up. A low threshold for evaluation of the cervical (C)-spine and ophthalmologic consultation must be maintained as well. There is a 2.3 to 10% concurrence rate between facial fractures and C-spine injuries. Blindness has been reported in as high as 3% of orbital fractures, and 12% of orbital fractures have concurrent ocular injuries.

Diagnosing facial fractures in pediatric patients may be difficult. Unless the patient is being evaluated immediately after the injury, swelling will have begun to set in. This may mask subtle physical findings and asymmetries. If a clear indication for operative intervention is not apparent, a delayed reevaluation after the swelling is resolved is necessary to appreciate the resultant deformity. Examination of the available patient photographs, dental records, and occlusal models is helpful.

A comprehensive craniofacial examination from the vertex to the clavicles should be completed for all patients. The patient must be rolled, and the entire face and scalp must be examined and palpated for lacerations, ecchymosis, swelling, hematomas, and step-offs. Lacerations should be irrigated and probed for fractures prior to closure. A complete examination includes otoscopic examination of the tympanic membrane, ophthalmologic evaluation, examination of the C-spine, and palpation of the neck for sub-Q subcutaneous emphysema and expanding hematoma. If concern is raised for airway stability, the patient should be intubated. This may be complicated by the presence of a C-collar and/or midface or mandibular instability. Pediatric anesthesia may be of assistance in complicated cases, and tracheostomy and cricothyrotomy are always means of establishing a definitive airway in an emergency setting. Further evaluation includes a full cranial nerve examination.

Signs of orbital trauma such as swelling, ecchymosis, subconjunctival hemorrhage, enophthalmos, exophthalmos, diplopia, limited extraocular mobility, altered visual acuity, irregularity or asymmetry of the pupil, a relative afferent pupillary defect (RAPD), and medial or lateral canthal malposition should be noted. Careful intra- and extranasal evaluations should be carried out. Palpation of fractured nasal bones may reveal irregularities, crepitus, or mobility. Loss of nasal projection may indicate a complex nasal bone and septal injury, or it may indicate the presence of a naso-orbito-ethmoid (NOE) fracture when the intracanal distance is increased. The nasal septum must be carefully evaluated for the presence of a septal hematoma, as failure to drain this may result in septal necrosis and a subsequent saddle-nose deformity.

Intraoral examination must also be carried out, with attention paid to dental trauma, intraoral lacerations, widened interdental distances, ecchymosis, and hematomas. These may indicate the presence of alveolar, maxillary, or mandibular fractures. Bimanual manipulation of the maxilla may indicate the presence of midface instability and a Le Fort fracture. A complaint of subjective malocclusion is sensitive but not specific for detecting fractures. This history is often unattainable in children and is therefore even less useful. A unilateral or anterior open bite may indicate the presence of condylar fracture. Trismus, or difficulty in opening the mouth, may be caused by a mandibular fracture, impingement by a depressed zygomaticomaxillary complex (ZMC) or zygomatic arch fracture, or swelling in the muscles of mastication from direct trauma. The mandibular condyle can be palpated by moving the jaw while placing one finger in the external auditory canal (EAC) and one finger in front of the ear. Laceration of the EAC on otoscopic examination may indicate a condylar fracture as well as a temporal bone fracture.

Radiographic Evaluation

Fine-cut (1–2 mm) maxillofacial computed tomographic (CT) scans have become the gold standard for assessing fractures of the craniofacial skeleton. Plain radiographs lack sensitivity, and adequate interpretation of these studies is becoming a lost art. The one possible exception to this is the pantomogram, or Panororex. This panoramic study provides a comprehensive, two-dimensional (2D) survey of the anatomy of the maxilla and mandible with low costs and radiation doses. It has comparable sensitivity to current CT scanners in identifying fractures of the mandible but has less inter-observer agreement when ruling out the presence of a fracture. Furthermore, it lacks the three-dimensional (3D) information obtainable from a CT study. These 3D reformats have become invaluable for characterizing fracture patterns and optimizing exposures and fixation strategies.

When caring for pediatric patients, surgeons must be thoughtful of the possible effects radiations have on the cognitive development of the growing brain and of the oncologic risk from such exposures. In a large retrospective study of the British National Health Service, obtaining two to three head CTs in patients younger than 15 years could lead to a threefold increase in the incidence of brain tumors. The absolute risk is less staggering. This number translates to only 1 additional tumor for every 10,000 scans performed. This risk is small, and although it must be considered, it should not dissuade a surgeon from obtaining studies needed for good clinical management.

7.4 Cranial/Frontal Sinus Fractures

7.4.1 Diagnosis

With the increased craniofacial proportions observed in small children, skull fractures are common and diminish in prevalence with age. In the authors' series, they represented the second most common fracture type in children younger than 6 years. These injuries typically present as linear fractures that may extend into the orbital roof, skull base, nasal capsule, or zygoma in oblique trajectories. In children younger than 5

years, accompanying dural lacerations may result in a growing skull fracture, as pulsations cause fracture widening and herniation of cranial contents. This occurs in less than 1% of skull fractures. The parietal bone is the most common location, but it can occur in the frontal bone or orbital roof. It then results in frontal swelling or pulsatile exophthalmos (► Fig. 7.2).

Patients with frontal sinus, cribriform plate, and other cranial base fractures should be interrogated for cerebrospinal fluid (CSF) leaks. In compliant patients with a cleared C-spine, they should be asked to sit forward and observed for clear-liquid draining from the nose. They may complain of salty fluid in the pharynx. Edema, rhinorrhea, epistaxis, an unstable spine, or altered mental status may complicate this assessment. Cerebrospinal fluid will demonstrate a "ring sign" when bloody drainage is blotted on an absorbent surface. Clear CSF will migrate further than blood when absorbed. In addition, when available, a β -2 transferrin assay is highly sensitive in differentiating CSF from other body fluids. Otorrhea may indicate a temporal bone fracture with CSF leak. Otoscopic examination is necessary to detect this and other signs of temporal bone trauma such as hemotympanum and laceration of the EAC.

7.4.2 Nonoperative Management

The goals of managing cranial and frontal bone injuries are protection of the neurocranium, repair of dural injuries, resolution of CSF leak, prevention of infection, and correction of aesthetic/contour deformities. In the absence of significant displacement or step-off, nasofrontal outflow obstruction, pneumocephalus indicating dural injury, gross contamination, or underlying neurotrauma, nonoperative management is preferred. Owing to the rapid cranial expansion seen in the first 3 years of life, significant remodeling can be expected, and depressed skull fractures are more likely to be successfully managed nonoperatively in small children. Most CSF leaks will resolve spontaneously. Typically, a course of bed rest and head elevation is adequate. If this fails, a period of CSF diversion via a lumbar drain or external ventricular drain is indicated before surgery.

In patients with a frontal sinus, patency of the nasofrontal duct is paramount. Delayed abscess and mucocele may develop years after an injury if this structure is injured. In the setting of a patent duct and minimally displaced fractures, nonoperative management is warranted, but serial examination and repeat imaging 3 to 5 years later are necessary. Serial follow-up is necessary in younger patients with frontal fractures to clinically rule out the development of a growing skull fracture.

7.4.3 Operative Management

Surgical treatment of skull fractures in children is rarely needed. In the senior author's series of isolated cranial fractures, 86% were managed nonoperatively; 7.5% required neurosurgical interventions such as external ventricular drain (EVD) placement, hematoma evacuation, and decompressive craniectomy; 6.5% underwent operative repair of the fracture, open reduction internal fixation (ORIF), frontal sinus repair, and fracture debridement. Frontal bone injuries were more likely to necessitate operative intervention due to the presence of the frontal sinus, orbital complications due to orbital roof involvement, and greater cosmetic sensitivity in this location.



Fig. 7.2 Growing skull fracture. (a) A 10-year-old child with a growing skull fracture of the left forehead and orbit after trauma. (b) Three-dimensional CT scan revealing the fracture. (c) Intraoperative view of the growing skull fracture. (d) Reconstructed fracture. (e) One-year follow-up, with normal forehead and orbital morphology.

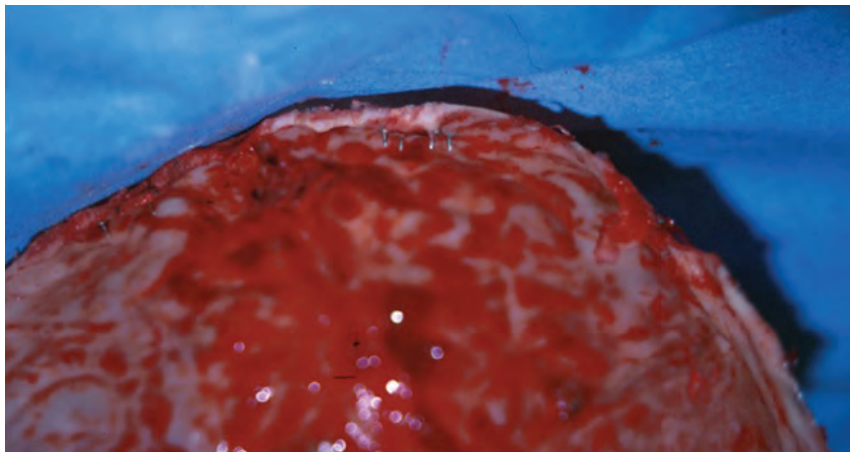


Fig. 7.3 Transcranial migration of metallic hardware. The use of metallic implants should be avoided for osteosynthesis in the growing cranium. In addition to growth at the sutures, the cranium grows through concurrent appositional growth, with deposition of new bone on the external cortex and resorption of the internal cortex. This results in transmigration of permanent implants into the skull. Hardware may become imbedded in the dura, brain parenchyma, or venous sinuses. The use of suture fixation or absorbable hardware made with poly(glycolic acid) (PGA) or poly(lactic-co-glycolic acid) (PLGA) is suggested.

Frontal bone injuries can be approached directly through extensive local lacerations or via a coronal approach. Any child with remaining cranial growth potential must have fractures fixated with absorbable hardware, as metallic implants are known to translocate through the skull with future appositional growth (► Fig. 7.3). These plates and screws may become imbedded in dura, brain parenchyma, or dural sinuses and present significant potential morbidity. Growing skull fractures are repaired with craniotomy to allow access and repair of the

underlying dural injury. Reconstruction then requires reconstitution of the normal anatomy, with cortical bone grafts in a configuration that places a contiguous piece of cortical bone overlying this repair.

When a frontal sinus is present, the algorithm developed by Rodriguez and Manson represents the most evidence-based approach to managing these injuries. The essential decision-making factor is the presence of trauma and obstruction of the nasofrontal outflow complex. When the nasofrontal outflow

complex is uninjured, displaced anterior wall fractures may be managed with simple ORIF, as long as duct patency can be documented intraoperatively. If the drainage pathway is either obstructed or likely to become obstructed due to scarring from an injury, the sinus must be defunctionalized. This requires complete removal of the sinus mucosa by burring the entire cortical surface, as epithelial cells are known to be present along draining venous sinuses in the bone and can serve as a tissue source for reepithelialization. The sinus duct should be filled with a well-fitting cortical bone plug to promote ossification. Management of the remaining sinus depends on the fracture patterns. In the setting of significantly displaced posterior table fractures, it is best to cranialize the sinus. The posterior table is removed, dural injuries are repaired, the obliterated sinus duct is covered with vascularized tissue such as a pericranial or galeal frontalis flap, and the cranial contents are allowed to expand into the frontal sinus space. When the posterior sinus wall is intact, the sinus may be managed by obliteration, in which the defunctionalized sinus is filled with various graft materials, including fat, fascia, and bone. Osteoneogenesis—a technique in which the duct is obliterated, the sinus is demucosalized, and the empty sinus is allowed to fill with bone and fibrous tissue over time—has also been described. Higher complication rates have been shown with the use of fat to fill the sinus dead space and with osteoneogenesis, and the authors' preferred technique is to utilize bone, harvested locally from the cranium or the iliac crest. In a patient with a developing frontal sinus, the authors have demonstrated that even significantly displaced anterior table fractures have the potential of remodeling with time and continued pneumatization of the frontal sinus (► Fig. 7.4).

7.5 Orbital, Zygomaticomaxillary, and Naso-Orbito-Ethmoid Fractures

7.5.1 Diagnosis

The orbit represents the intersection between the cranium and the facial skeleton. In the authors' series, orbital fractures represent the most common fractures in pediatric patients of all ages, representing 29.8% of fractures. In children aged 5 years or younger, orbital fractures represented 56.4% of fractures. This is

likely due to the high cephalic-to-facial ratio and differences in midface anatomy, discussed previously. According to the authors' findings, as children aged, the incidence of orbital fractures remained high but decreased as children began to suffer more classic adult patterns of injury.

Because of the propensity for various fracture patterns to transgress the orbit in pediatric injuries, the senior author has described a classification system for pediatric orbital fractures. Type 1 injuries represent pure orbital fractures, without involvement of adjacent structures. Included within this group are isolated orbital roof fractures, which are more common in early childhood and are replaced by floor fractures as the child matures. Type 2 fractures represent oblique craniofacial fractures involving the frontal bone, orbit, and, possibly, midface. Finally, type 3 fractures represent more common adult patterns of complex midface trauma, including ZMC and NOE fractures.

Orbital examination is critical in all patients with facial fractures. The authors recommend an ophthalmologic consultation for all patients with orbital fractures to rule out ocular trauma and document a baseline visual acuity. This is especially important for nonverbal patients and in any patient for whom operative intervention is considered. With fractures extending toward the superior orbital fissure, ptosis and ophthalmoplegia may indicate a diagnosis of superior orbital fissure syndrome (SOFS), with trauma to the third, fourth, and sixth cranial nerves. Diminished visual acuity and an RAPD indicate traumatic optic neuropathy (TON). If SOFS and TON are present together, the correct diagnosis is orbital apex syndrome. Involvement of ophthalmology in these cases is critical, as vision is threatened and management is nuanced. Surgical decompression may be indicated if an offending anatomic lesion is identified. The use of mega-dose steroids is controversial, with little evidence demonstrating their efficacy. Their use should be on an individualized basis in the setting of an interdisciplinary approach.

Visual inspection of the eyelids for lacerations and injuries to the lacrimal apparatus is important. Lacerations of the eyelid medial to the punctum indicate a lacrimal disruption, until proven otherwise. Integrity of the medial canthal tendon must be confirmed with a bowstring test. The bowstring test is performed by the examiner by placing a finger over the insertion of the medial canthal tendon while exerting lateral traction on the eyelids. Resistance to lateral traction and an intact medial tendon insertion will be appreciated. A mobile medial canthus, or asymmetric canthal position, relative to the midline,

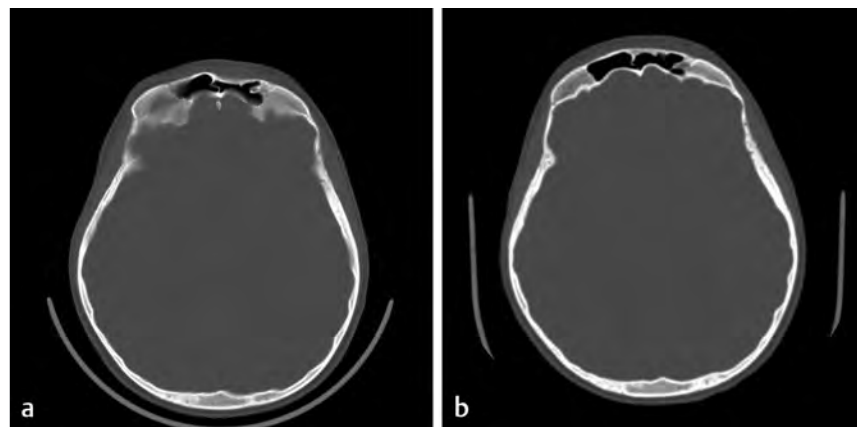


Fig. 7.4 Anterior table frontal sinus fracture remodeling. (a) A patient with depressed anterior table frontal sinus fracture, without a posterior table fracture or injury of the nasofrontal outflow tract, who presented for plastic surgical evaluation 4 weeks after the injury. The family wished to avoid surgery, and a conservative follow-up course was adopted. (b) One-year follow-up CT scan revealed remodeling of the anterior table without contour irregularity and the absence of retained fluid in the sinus. An additional follow-up with repeat imaging 5 years after the injury was recommended to ensure that the patient had a “safe sinus.”

indicates either a canthal avulsion or an NOE fracture. Asymmetry of the lateral canthus with inferior or posterior displacement may indicate a displaced fracture of the zygoma. Note should be made of subconjunctival hemorrhage, as this tissue plane is continuous with the periorbital and, in the setting of significant trauma, is diagnostic of a periorbital fracture, until proven otherwise. The surgeon should also look for signs of enophthalmos, a finding that will often be masked by swelling in the acute setting. After resolution of swelling, narrowing of the palpebral fissure, asymmetry of the supratarsal crease, and deprojection of the globe are diagnostic. Exophthalmos may also be present in the setting of an impacted zygoma, fronto-temporal orbital fracture, or orbital roof “blow-in” fracture. Pulsatile exophthalmos can develop in traumatic carotid cavernous fistulas and delayed presentation of a growing skull fracture.

Restricted extraocular motility may have multiple causes, including neurologic lesions such as SOFS; edema and hematoma in an extraocular muscle; globe malposition, altering the functional advantage of the extraocular muscle; or muscle entrapment. Extraocular muscle entrapment may be indicated by radiographic findings on a CT scan (► Fig. 7.5) and is diagnosed clinically. It is rare for large orbital defects to cause muscle entrapment, though septae connecting the periorbital to the extraocular muscles may become snared on bony edges. This mechanism is more common in adults. In young children, greenstick, linear trapdoor fractures of the orbital floor have a propensity to close and trap herniated periorbital fat or muscle, once the offending compressive force is removed from the orbit. On physical examination, an individual will have diplopia and pain on superior gaze and a firm stopping point for ocular excursion confirmed with a forced duction test. These patients typically present with increased parasympathetic tone due to an ocular–cardiac reflex and may be bradycardic and nauseated. This diagnosis is a true emergency, as the entrapped muscle may easily become ischemic, resulting in potential scarring and impaired excursion in the future. This can cause disabling diplopia.

ZMC fractures typically present with displacement of the lateral canthus, flattening of the malar prominence, trismus from

zygomatic arch impingement on the temporalis muscle, and subconjunctival and buccal sulcus hematomas. Exophthalmos may be present in cases where significant impaction is present; and, enophthalmos may develop postoperatively if reduction of the fracture is performed without recognizing that reduction of the impacted segment often results in a significant orbital floor defect.

NOE fractures are the least common injury in the pediatric facial skeleton. Fractures of the nasofrontal buttress, nasomaxillary buttress, and medial orbital wall can result in traumatic telecanthus and loss of dorsal nasal support. The medial orbital wall segment, or “central fragment,” bears the insertion of the medial canthus and if displaced, results in traumatic telecanthus. Fractures are classified based on the degree of comminution present. Type 1 fractures include a single fragment; type 2 fractures are comminuted but the canthal tendon remains attached to a substantial bony fragment, which can be reduced and fixed in position; and type 3 fractures are severely comminuted with an avulsed canthal attachment. Displacement of bony segments may result in swelling or trauma to the central lacrimal apparatus. Typically, this is corrected with proper reduction of the fractures.

7.5.2 Nonoperative Management

Indications for operative treatment of orbital fractures are less common in pediatric patients. In adults, clear indications such as blow out fractures with a greater than 1 cm² defect, greater than 50% orbital wall displacement, or bony impingement causing SOFS or TON are clear-cut. The authors believe that the periorbital and supporting structures of the pediatric globe are more resilient than those of the adult. Pediatric patients, with even large bony defects following trauma, often do not develop enophthalmos, vertical orbital dystopia (VOD), or diplopia. For this reason, isolated pediatric orbital fractures are best observed in the absence of acute enophthalmos, VOD, or muscle entrapment.

In the senior author’s series, type 1 pure orbital fractures were managed nonoperatively in 88% of cases. Oblique

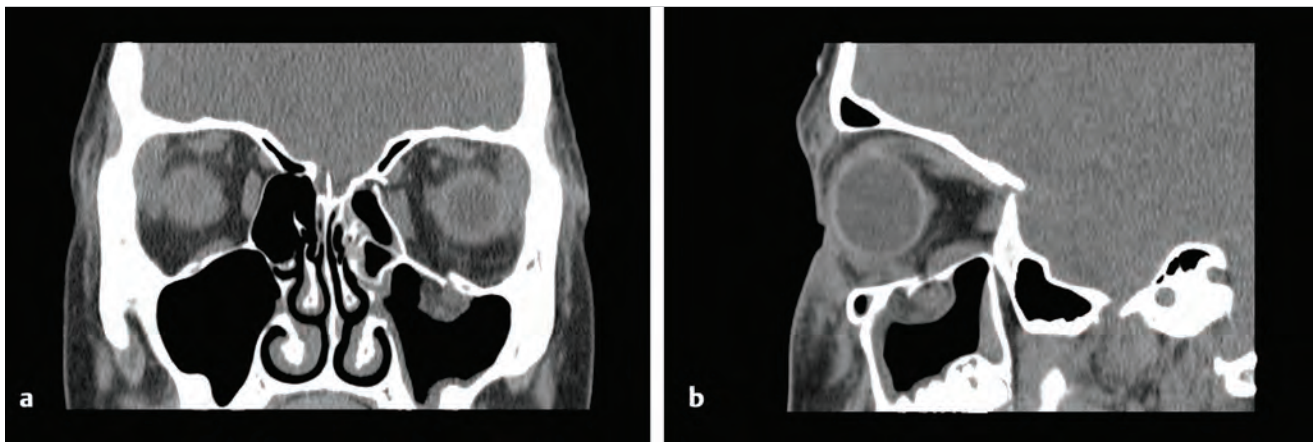


Fig. 7.5 Inferior rectus entrapment in an orbital floor “trap door” fracture. (a) Coronal and (b) sagittal CT images demonstrating entrapment of the inferior rectus muscle in a linear, nondisplaced, “trap door” fracture of the orbital floor. Findings of CT scan may be highly suggestive and illustrate the mechanism of entrapment well, but the diagnosis of entrapment must be made by clinical examination. Patients will typically have diplopia, limited excursion of the affected eye when gazing upward, and pain with eye motion; they may also have nausea and bradycardia. A forced duction test will reveal a firm stop preventing upward eye mobility.

craniofacial injuries, or type 2 fractures, were conservatively followed unless a clear operative indication was encountered; and, only 17% ultimately required intervention. Type 3 injuries, which represent impure orbital fractures involving the surrounding skeleton (inferior orbital rim, ZMC, or NOE), were more likely to require operation. In the author's series, 72% of impure orbital fractures underwent operative repair. These were the least common injuries, representing 25.9% of cases.

7.5.3 Operative Management

Ophthalmologic evaluation is recommended prior to operative treatment of periorbital facial fractures. In the setting of penetrating globe trauma, retinal pathology, TON, or traumatic glaucoma a delay in surgery may be warranted. This decision should be made in conjunction with a trained ophthalmologist.

Acute orbital muscular entrapment with a positive forced duction test is an operative emergency in the pediatric population. Often, muscle and/or fat is caught in the fracture line. This tissue may become ischemic and the best results are achieved with prompt exploration and release. Other indications for operative treatment include acute enophthalmos, presentation of clinically significant enophthalmos after swelling resolution, vertical orbital dystopia, and impingement from bony fragments.

Access to the orbital floor can easily be achieved through a transconjunctival approach in either a preseptal or a retroseptal plane, with limited risk of ectropion and lid malposition. If needed, this approach can be augmented with a transcaruncular incision to achieve exposure of the medial orbit and/or a lateral canthotomy to gain access to the lateral orbital wall. Reconstruction of the orbital floor and walls requires subperiosteal dissection 360 degrees around the perimeter of the defect. Malpositioned orbital contents are returned to the orbit, and an implant is placed to partition the two cavities. This implant must sit on all edges of the defect, including the posterior margin so that it does not settle into the sinus and cause postoperative enophthalmos. While dissecting to reach the posterior ledge of the defect, the surgeon must remember that the optic nerve is 40 to 45 mm from the orbital rim; however, these dimensions will be smaller in younger patients. Orbital defects may be reconstructed with autologous split calvarial bone, or alloplastic materials such as absorbable mesh, titanium, and polyethylene. These constructs should be rigidly fixed to the orbital rim to prevent implant migration. Autologous bone reconstruction is ideal, but one must weigh the morbidity of a graft donor site against the likelihood of infection and implant migration.

ZMC fractures are rare in children due to the lack of an aerated maxillary sinus. When they occur, they are likely to be significantly displaced and require operative reduction. Some authors have described open reduction via a buccal sulcus incision alone, with fixation at the orbital rim and zygomaticomaxillary buttress. However, the authors feel that this is inadequate, as the best location to assess reduction of the fracture is at the lateral orbital wall along the zygomaticosphenoid suture. Exposure of the orbital rim and floor can be achieved via a transconjunctival approach, but a cantholysis is necessary to assess the lateral rim through this incision. Alternatives include augmenting the transconjunctival approach with a lateral blepharoplasty incision or substituting a subciliary approach with lateral canthal extension in a crow's foot crease. Care must be taken to

avoid extending the lid incision into the temporal area where the scar will be more noticeable. Some authors have reported that the subciliary approach carries a slightly higher risk of ectropion than the transconjunctival approach; however, the senior author has found that a carefully planned and executed subciliary approach with lateral canthal extension into a crow's foot crease, combined with robust soft tissue resuspension, can be an acceptable approach.

Adequate reduction of the ZMC fracture is achieved at the zygomaticofrontal suture, infraorbital rim, and zygomaticomaxillary buttress. Typically, reduction and fixation are achieved in that order. Ensuring that the lateral orbital wall is aligned is the best way to confirm that the fracture is reduced in all three dimensions of space. Exploration of the orbital floor is necessary to confirm that the configuration of the orbital floor has not been altered after reduction and floor reconstruction is not warranted. Exposure through these three combined approaches necessitates wide undermining of the entire malar prominence. Soft tissues must be resuspended to the orbital rim to prevent cheek and lid ptosis.

Significant NOE fractures are characterized by posterior and lateral displacement of the nasal bone and medial orbital walls. Reduction typically requires a combination of coronal, transconjunctival, and buccal sulcus incisions. Anatomic reduction of a type 1 or 2 fracture should place the canthal tendon in a proper location. Type 3 fractures often need bone grafting and transnasal wiring with a posterior-superior vector to correct the traumatic telecanthus. In severe injuries, overcorrection of the medial canthus and soft tissue adaptation with bolsters are often needed. Despite anatomic reduction, handling the soft tissue often determines the result. Bone grafts may be needed to reconstruct the medial orbital wall and the nasal dorsum.

Children must be observed postoperatively following orbital surgery, with visual acuity documented. Head elevation, cold compresses, and steroids may help alleviate swelling and chemosis. After significant manipulation of the lower eyelid, the surgeon should consider placing a Frost stitch to support the lower eyelid during early healing and edema resolution and to minimize eyelid malposition. Any change in visual acuity or unexplained pain should prompt radiographic evaluation. In the setting of an orbital fracture and reconstruction, a retrobulbar hemorrhage resulting in an orbital compartment syndrome is exceptionally rare; however, it should always be considered.

7.6 Nasal Fractures

7.6.1 Diagnosis

Nasal fractures are the second most common pediatric facial fracture. Their incidence increases with age. The diagnosis can often be made clinically. Radiographs are often unnecessary; however, with significant injuries, a CT scan is helpful in delineating pathologic anatomy, particularly of the nasal septum. External inspection and endonasal examination for nasal deviation, swelling, lacerations, and septal hematoma are key.

7.6.2 Nonoperative Management

Most practitioners are reticent to perform open manipulation or resection of the septum, or osteotomies of the nasal bones,

on growing patients. For this reason, corrective septorhinoplasty for secondary nasal deformities is typically deferred until midface skeletal maturity. Operative treatment should be considered for children with nasal fracture, in the form of a closed reduction and external fixation. Patients with nondisplaced injuries require no intervention other than limiting activity for 6 weeks to protect the nose. If significant swelling is present, the patient should be reevaluated after several days to determine if a deformity is present. If displacement is noted, all medically stable children benefit from a closed reduction under anesthesia, to at least lessen the deformity, if not correct it, as they will not be candidates for formal septorhinoplasty until they have reached the age of puberty and midface skeletal maturity.

7.6.3 Operative Management

Initial treatment of nasal fractures consists of closed reduction under anesthesia, with a protected airway. An elevator is placed intranasally to elevate depressed nasal bones, and manual manipulation can straighten the dorsum and improve symmetry. Ash forceps can be used to straighten a fractured septum. The nasal complex is immobilized with tape and an external splint, whereas Doyle splints are used to stabilize the septum. Nasal packing may be used, when required, to maintain reduction of a freely mobile and collapsed nasal bone. Antibiotics are needed while internal splints are in place. Doyle splints and nasal packing can be removed in 5 to 7 days, and external splints can be removed after 1 to 2 weeks.

Nasal and septal fractures may result in development of a crooked nose, dorsal hump, and residual deformities—commonly seen after closed reduction and external fixation. Residual septal distortion or scarring may contribute to functional nasal obstruction. These deformities can be addressed in a delayed fashion by using traditional septorhinoplasty techniques, but these techniques should be deferred until adolescence.

7.7 Maxillary and Dentoalveolar Fractures

7.7.1 Diagnosis

Maxillary fractures are less common in young children and become more prominent as the maxillary sinus develops and the permanent dentition erupts. At younger ages, impacts are absorbed by the cranium and orbital region. Blows to the midface are dissipated by the flexible bone and deflected to the alveolar ridge. Palatal splits are more common due to a patent midpalatal suture.

Important attention must be paid to diagnosing malocclusion in patients with maxillary trauma. This can be difficult in children with mixed dentition (6–12 years old). The presence of mamelons indicates a tooth that has never been in occlusion. When available, dental records and occlusal models are invaluable in determining the preoperative occlusion. These can be used to fabricate an occlusal splint for use in the operating room. Bimanual manipulation of the midface is key to determine if a mobile Le Fort segment is present, which requires fixation.

Attention must be paid to the dentition. Mobile, missing, and fractured teeth must be noted. Diastasis between teeth may indicate a palatal fracture. A pediatric dentist is a valuable ally in evaluating and triaging injuries to the dentition and alveolar segment.

7.7.2 Nonoperative Management

The growing maxilla has a great deal of plasticity with growth and dental eruption. Orthodontics can correct many minor occlusal irregularities caused by fractures. Open reduction and internal fixation also places unerupted dental follicles within the maxilla at risk of injury. For this reason, it is often best to accept minor malocclusions in young children and manage them nonoperatively. Dental eruption is typically completed at around 12 years of age, with the exception of the third molars. When patients are treated conservatively, jaw rest with a pureed diet until the fracture has healed is prescribed. Regular follow-up visits to gauge compliance and to monitor for worsening malocclusion are indicated.

7.7.3 Operative Management

Mobile dentition involved in dental alveolar fractures should be stabilized using arch bars, occlusal splints, or bonding to the adjacent dentition. Arch bars are helpful for maxillomandibular fixation (MMF), but placement can be difficult in children. However, the authors have reported the successful use of arch bars in primary and mixed dentition. The primary dentition has short roots and conical crowns that make it difficult to cinch wires tightly. Crowding in mixed dentition can also complicate their application. More inventive means of MMF are often needed, such as Risdon's cables and pyriform drop wires combined with circummandibular wires. Palatal fractures can often be handled conservatively with a palatal splint, with or without MMF following reduction. Open fixation is sometimes warranted. When MMF is applied in children, the course is typically shortened due to rapid fracture healing and the propensity for ankylosis of the temporomandibular joint (TMJ). One week or less of rigid MMF can be followed by 2 to 3 weeks of elastics, with close observation for recurrent malocclusion. When ORIF is indicated, standard exposure and fixation of the zygomaticomaxillary and nasomaxillary buttresses are indicated. Care must be taken to avoid traumatizing unerupted tooth follicles. In addition, if greenstick fractures prevent reduction, the fracture may need to be completed to allow full mobilization and placement of the segments in occlusion before plating.

7.8 Mandible Fractures

7.8.1 Diagnosis

Trismus, intraoral swelling, hematoma, dental trauma, and malocclusion should warn the clinician of a possible mandible fracture. Condylar and condylar head fractures represent the most common location for pediatric fractures. The condyle is hypervascular and less mineralized than the adult counterpart, making it prone to fracture. Falls with a blow to the chin are a common mechanism, and injuries such as this are often the presumed etiology of mandibular asymmetry and TMJ

ankylosis. Distinct patterns of malocclusion are commonly witnessed with subcondylar fractures and should be identified. In unilateral injuries, the fragments commonly override, causing shortening of the condylar unit and premature dental contact ipsilateral to the injury. An open bite is observed on the contralateral side. Bilateral injuries will cause an anterior open bite through the same mechanism.

7.8.2 Nonoperative Management

The mandibular condyle possesses a remarkable propensity for remodeling and growth in the first decade of life. Prolonged immobilization may predispose children for the development of TMJ ankylosis. The presence of dental follicles and the inferior displacement of the inferior alveolar nerve result in limited real estate for the placement of fixation hardware. For these reasons, conservative management of pediatric mandible fractures is often favored. Nondisplaced or minimally displaced fractures in young children can often be successfully managed with immobilization by using a jaw bra or cervical collar, along with a liquid diet. Minor malocclusions may be corrected later with orthodontic manipulation.

Owing to the risk of ankylosis, intracapsular condylar head injuries should not be immobilized. Typically, a short period of mandibular rest with a liquid diet is followed by chewing as the physical therapy to maintain mobility. In fact, the presence of a condylar fracture should be considered an indication for ORIF of concurrent fractures elsewhere in the mandible to avoid the need for MMF. The most common combined fracture of the pediatric mandible includes the symphysis/parasymphysis along with a condylar (head or neck) fracture. The authors recommend the placement of arch bars, an ORIF of the symphysis/parasymphysis fracture, and the postoperative use of elastics and physical therapy of the TMJ as a treatment regimen. Consultation with a pediatric dentist and/or craniofacial orthodontist is also very helpful.

7.8.3 Operative Management

When managing pediatric mandible fractures, the goal is to restore occlusion while limiting injury to dental follicles or future growth. Whenever possible, intraoral approaches or pre-existing lacerations should be utilized for access while remaining cognizant of fracture fragments' blood supply. As in maxillary trauma, placing children in MMF may be difficult, and alternative methods may be needed.

When required, the period of MMF is abbreviated in pediatric patients. Often, unilateral subcondylar fractures can be managed with a short course of mandibular rest in MMF (5–7 days), followed by 2 to 3 weeks of contralateral guiding elastics and close observation for recurrent open bite. This contralateral force is able to overcome the masticatory muscles on the side of the injury, preventing overclosure of the fractured side and a resulting contralateral open bite. Treatment algorithms for bilateral injuries are more controversial. When posterior height is lost with a resultant open bite, the youngest children will require a course of MMF. In older children approaching skeletal maturity, this scenario represents a reasonable indication for ORIF of one or both sides, followed by MMF for the management of an unrepaired condyle. Other indications for ORIF of

the condyle include foreign body in the temporomandibular joint (TMJ), failure of closed reduction to obtain normal occlusion, and displacement of the condyle into the middle cranial fossa.

Displaced fractures of the angle, body, symphysis, and parasymphysis require ORIF. Angle fractures can be managed with a single superior border plate (Champy's technique). Accepted principles for the management of other fracture types include an inferior border plate with bicortical screws, along with a superior tension band. In children, hardware must be placed at the inferior border and monocortical screws should be used to protect tooth buds. Bicortical screws may be used after the secondary teeth have erupted. A bridle wire, arch bar, or small plate with monocortical screws may be used as a tension band. In small children, an inferior border interosseous wire, bridle wire, and a brief course of MMF may be substituted. Metallic fixation should be considered for later removal to prevent any potential growth restriction. When approaching the symphysis and parasymphysis, preservation of a cuff of mentalis muscle attached to the bone is necessary for resuspension during closure to prevent chin ptosis.

7.9 Panface Fractures

Open reduction and internal fixation of panfacial fractures should proceed in a systematic manner. It is necessary to expose all fractures through multiple approaches before initiating the sequence of fixation. All buttresses must be reconstructed, with attention paid to restoring the width, height, and projection of the face. Fracture fixation must start from a stable, uninjured point. The sequence of fixation may proceed from the cranium downward or from the mandible upward. Similarly, fixation may begin centrally and progress laterally or may begin laterally and progress centrally. The process must be sequential and not haphazard. Cases with maxillary injuries and mandibular condyle fractures represent an indication for ORIF of the condyle to reestablish facial height. Similarly, in a true panface fracture, ORIF of the zygomatic arch via a coronal approach may serve as a reliable landmark to establish facial width when all other buttresses are fractured and unreliable.

7.10 Outcomes

Owing to concern for altered growth, adequately defining outcomes following pediatric facial fractures remains an elusive goal. Reported complication rates vary from 2.1 to 32.2%. Serial clinical and radiographic follow-up until skeletal maturity is needed to completely evaluate the outcomes of a treatment protocol. With this in mind, the authors have proposed the following classification scheme for outcomes following pediatric facial fractures to facilitate thoughtful characterization and study of these events (► Table 7.1). Type 1 adverse outcomes directly result from the injury (i.e., blindness after orbital fracture). Type 2 adverse outcomes directly result from the treatment (surgical or nonoperative) of the fracture (i.e., ectropion after subciliary approach to an orbital fracture). Type 3 adverse outcomes result from interaction between the injury, its treatment, and subsequent growth and development (i.e., late mandibular asymmetry). The true incidence and causative factors

Table 7.1 Classification system for adverse outcomes in pediatric facial fractures

	Definition	Examples
Type 1	Adverse outcomes related to the injury	Blindness resulting from an injury or tooth loss
Type 2	Adverse outcomes resulting from the treatment of the fracture	Ectropion following subciliary approach, hardware failure, and malocclusion after open reduction internal fixation
Type 3	Adverse outcomes resulting from a combination of the injury, its treatment, and subsequent altered growth and development	Progressive hypoplasia, facial asymmetry, or failure of dental eruption

leading to abnormal posttraumatic growth remain somewhat elusive, and the role that hardware plays is similarly unclear. If metallic implants are used in treatment, subsequent removal should be considered, especially in younger patients and in cases where dental eruption is impeded by the hardware.

7.11 Conclusion

The neonatal craniofacial skeleton is dramatically different from its adult counterpart. Growth and development result in an amazing transformation during the first two decades of life. When treating a pediatric patient with facial trauma, the fourth dimension must be considered in every treatment decision. The goal is to ultimately recreate normal form and function, but the surgeon must recognize that the injury and any subsequent intervention may alter the course of normal growth. Conservative treatment strategies are often the best, but when surgery is indicated, it must be done with extensive knowledge of the different anatomy and growth processes present in the patient.

7.12 Key Points

- Variations in anatomy result in different fracture patterns in children. Young children exhibit more fractures of the cranium and orbit. Complex facial trauma takes the form of oblique craniofacial fractures rather than fractures with Le Fort pattern. As the craniofacial skeleton matures, more mature patterns of injuries are seen in adolescents.
- With remaining growth and dental eruption, the pediatric facial skeleton is amazingly adaptive. It is often best to accept minor malocclusions or minimally displaced fractures as future growth, and remodeling may correct these.

- When operative intervention is warranted, care must be taken to avoid trauma to the growth sites and dental follicles.

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8 Cranioplasty

Arin K. Greene and Gary F. Rogers

Summary

The two most important variables to be considered for cranioplasty are whether the problem is related to contour (onlay) or protection of the brain (inlay) and the age of the patient. Because onlay cranioplasty generally is performed to improve an aesthetic deformity, we prefer to intervene when children are older and use an alloplastic material because there is less resorption compared to bone grafts. We favor porous polyethylene because it allows fibrovascular ingrowth and is easy to contour. Large full-thickness cranial defects necessitate closure to protect the brain. We allow spontaneous ossification to occur during infancy and intervene after this time. Particulate bone grafting/exchange cranioplasty is our first-line technique in children and adults because the bone grows with the child and completely osseointegrates. In adults with significant morbidities we will consider inlay cranioplasty using alloplastic materials to limit the extent of the operation

Keywords: cranium, cranioplasty, grafts, alloplastic, osseointegrate, fibrovascular

8.1 Introduction

Cranioplasty involves either the augmentation (onlay) or replacement (inlay) of cranial bone. Substances used for cranioplasty include (1) autologous bones (typically cranium, iliac crest, and rib) and (2) alloplastic substances (hydroxyapatite, demineralized bone, methylmethacrylate, porous polyethylene, and titanium). Advantages of autologous bone grafts are that they completely osseointegrate and thus provide strength and durability and have minimal risk of infection. Disadvantages of autologous grafts are limited availability, increased operating room time, and donor-site morbidity.

Alloplastic materials have three advantages: (1) supply is abundant, (2) there is no donor site, and (3) operative time may be shortened compared with autogenous techniques. However, these substances are not osteogenic (capable of new bone formation) and tend to be expensive. Demineralized bone is osteoinductive (able to induce transformation of undifferentiated mesenchymal cells to osteoblasts). The remaining substrates are either osteoconductive and provide a scaffold for bony ingrowth (e.g., hydroxyapatite and porous polyethylene) or have no biological activity (e.g., methylmethacrylate and titanium). Most bioactive alloplastic substances undergo incomplete revascularization and osseointegration and are susceptible to latent infection, foreign body reaction, displacement, and breakdown. The major variables that determine which material is used for cranioplasty are (1) whether the reconstruction is onlay or inlay and (2) the age of the patient.

8.2 Diagnosis

Most partial and full-thickness cranial defects can be diagnosed by physical examination. Large areas are noticed visually,

especially if they involve a non-hair-bearing region. Moderate to large bony deficiencies can be palpated. Generally, patients who are planning on cranial reconstruction have imaging of their defect to facilitate planning of the procedure. Computed tomography (CT) is the most commonly used imaging modality because it best delineates the bone. CT subjects the patient to radiation and thus should be used sparingly. Although we typically obtain CT prior to an operation, we often do not obtain postoperative CT to assess the reconstruction if the procedure is successful clinically.

8.3 Nonoperative Treatment

Intervention for patients with osseous defects is not mandatory. Those with partial-thickness deficits may suffer psychosocial morbidity if the deficiency is causing a visible deformity. A concavity in a hair-bearing region can be camouflaged with a wig, hat, scarf, etc. Patients with frontal bone deformities may be able to grow their hair sufficiently to cover the abnormality.

Full-thickness defects less than 1 cm in an adult often heal spontaneously (critical-size defect). In children younger than 12 months, much larger areas of exposed dura may spontaneously ossify. Consequently, it is best to wait several months in adults with small full-thickness cranial defects or longer in infants with larger areas to allow as much spontaneous ossification as possible. Some individuals will no longer require intervention, and if cranioplasty is required, the area to reconstruct will be smaller.

Patients with small full-thickness defects often do not require cranioplasty to protect the brain from trauma. However, individuals with larger deficits are at a greater risk for brain injury from trauma. Helmeting can be used to protect the brain in patients awaiting cranioplasty (usually children who are most susceptible to incidental trauma).

8.4 Operative Treatment

8.4.1 Onlay Cranioplasty

Children

The primary indication for onlay cranioplasty is to correct a visual deformity to improve the patient's self-esteem. Because the cranium has achieved more than 80% of its growth by the age of 4 years, elective procedures to improve appearance should be done after this time. In addition, a child's self-esteem does not begin to form until approximately 4 years of age. Onlay cranioplasty is often performed when patients are older and verbalize that they are bothered by the deformity.

Although autologous bone grafts are preferred for full-thickness defects in children, their use for onlay cranioplasty in the pediatric population is uncommon. Because underlying osseous continuity exists, there is less concern that an alloplastic substance will restrict cranial growth or become dislodged with growth. Nevertheless, bone thickening will occur around the implant, and loss of contour over time is not uncommon. Some

surgeons, however, prefer to use autologous bone grafts for onlay cranioplasty in children. Split cranial bone is the best autologous material. When harvested before 5 years of age, split cranial bone yields limited bone thickness and can be challenging due to a poorly formed diploic space. In situ harvest is not generally recommended before 9 years of age. Split graft is primarily cortical and heals by osteoconduction. The recipient site should not be burred to punctate bleeding, because enhanced vascularization will increase resorption of the graft through greater osteoclast activity. Cancellous bone graft from the iliac crest and particulate bone graft are not indicated for onlay cranioplasty because they resorb. These types of grafts require immediate vascularization to survive, which does not occur when they are placed on top of cortical bone.

Drawbacks of split cranial bone for onlay cranioplasty are donor-site morbidity (e.g., injury to the dura, bleeding, and thinner bone at donor site) and resorption of the graft. Cortical grafts may undergo significant resorption when placed over bone, and thus, patients are at risk for an unfavorable outcome and repeat cranioplasty. Most surgeons prefer alloplastic materials for onlay cranioplasty (even in older children), because there is no donor-site morbidity and minimal resorption (► Fig. 8.1). However, nonautologous substances have a higher infection rate than bone grafts.

Adults

Because the cranium is almost fully grown by 5 years of age, concerns regarding dislodgement of alloplastic substances or restriction of growth are not as relevant after this time. Consequently, many surgeons prefer alloplastic substances instead of autologous bone grafts, because they shorten operative time, do not expose the patient to donor-site morbidity, and have minimal resorption. Although there are numerous alloplastic substances that are used for onlay cranioplasty, the most common are hydroxyapatite, methylmethacrylate, and porous polyethylene (Medpore).

Porous polyethylene is our preferred alloplastic material. It allows osseointegration/neovascularization at the periphery of the implant and is easily moldable to fit the partial-thickness defect. Although custom implants can be ordered based on CT images, contouring standard pieces to the defect is easy and less expensive. Bone substitutes (e.g., hydroxyapatite) allow some

osseointegration at their periphery but are typically harder to contour than porous polyethylene and are more susceptible to fracture and infection. Methylmethacrylate is not commonly used for onlay cranioplasty, because it does not osseointegrate and has a higher fracture and infection rates compared with other materials.

8.4.2 Inlay Cranioplasty

Children

Inlay cranioplasty in children is challenging, because there is limited autologous donor-site material, and split cranial graft harvest may be challenging in very young patients. Although the cranium has completed most of its growth by 5 years of age, most surgeons prefer to use autologous bone graft until adolescence because the graft will (1) fully osseointegrate and (2) not restrict growth or become dislodged as the cranium expands. Bone grafts undergo less resorption when used for inlay cranioplasty compared with the onlay position. Dural-mediated osteogenesis supersedes the resorptive effect of revascularization and leads to a gain in volume.

The cranium is the preferred donor site for cranioplasty, because it is in the same field as the defect, there is less donor-site morbidity, a large amount of material can be obtained, and the bone undergoes minimal resorption compared with iliac or rib grafts (► Fig. 8.2). Donor-site problems from rib graft harvest include contour irregularity, pleuritic pain, and pneumothorax. Iliac procurement causes discomfort and risks injury to the lateral femoral cutaneous nerve. In contrast to iliac or rib donor sites, which require a second incision, the cranium is already exposed during the procedure. The cranial donor site causes minor discomfort, and the scar is well concealed. Cranial bone can be harvested as a full-thickness, partial-thickness, or particulate graft. Although full-thickness bone is excellent for structural purposes, most surgeons use split cranial bone graft through the diploic space to obtain two pieces of mostly cortical bone—one to perform the reconstruction and the other to repair the donor site. Limitations of split cranial bone grafting include (1) the segments over the donor and recipient sites do not result in normal thickness, (2) the technique requires a well-developed diploic space, and (3) certain areas of the cranium (e.g., temporal bone) are not amenable due to a narrow or absent diploic space.

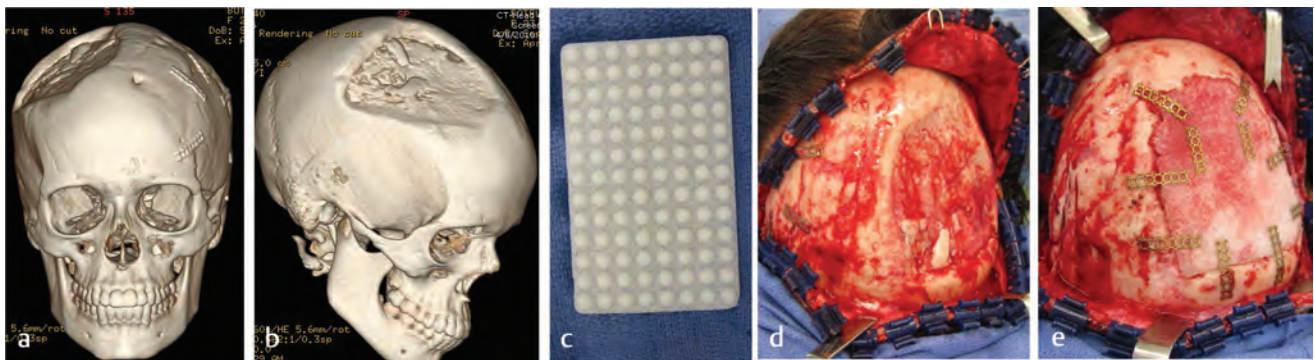


Fig. 8.1 An adolescent girl with a partial-thickness parietal defect managed by onlay cranioplasty using porous polyethylene. (a,b) Preoperative CT images. (c) Porous polyethylene. (d) Intraoperative view of defect (e) after onlay cranioplasty.

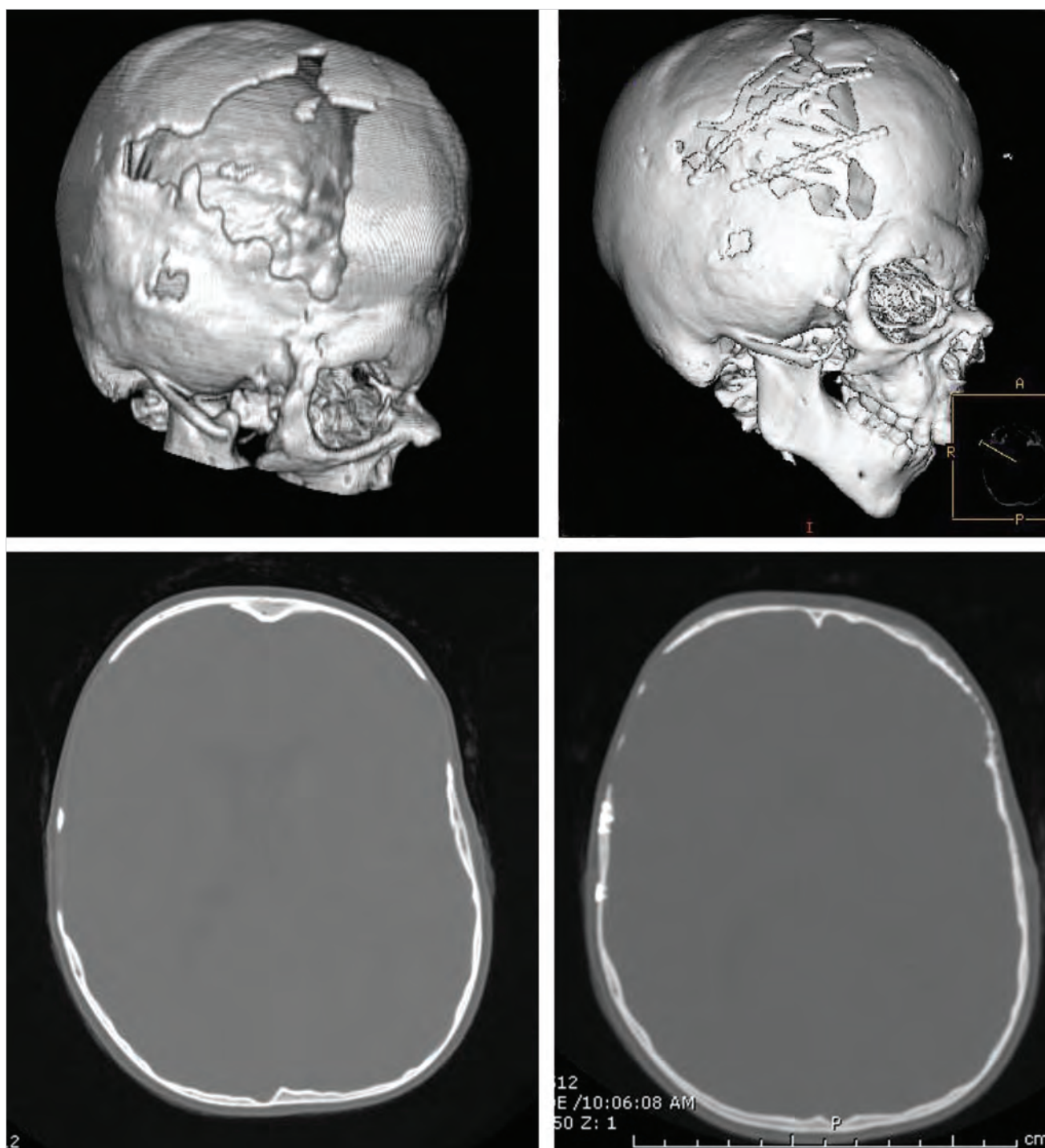


Fig. 8.2 Resorption of rib graft used for inlay cranioplasty. Preoperative cranial defect (left). Resorption of graft 2 years postoperatively (right).

Because the dura has the ability to spontaneously ossify until approximately 12 months of age, cranioplasty is generally not performed in infants, to allow the area become as small as possible. Our preferred method of inlay cranioplasty in infants and young children is cranial particulate bone grafting, because this technique does not require a diploic space for harvest and is simpler and safer than split cranial bone graft (► Fig. 8.3). A 16-mm Hudson's brace and D'Errico's craniotomy bit (Codman & Shurtleff, Inc) are used to harvest corticocancellous bone from

any area of the cranium. The manual drill prevents thermal injury and ensures maximal graft viability. The material is mixed with blood, applied to the dura that is at least 0.5-cm thick (or to match the thickness of the surrounding bone), and covered with fibrin glue. Particulate graft heals by osteogenesis and works best when placed over normal or minimally scarred dura. Over time, the graft becomes indistinguishable from the surrounding bone and can even form a diploic space (► Fig. 8.4). An advantage of particulate graft is that the donor sites are of partial

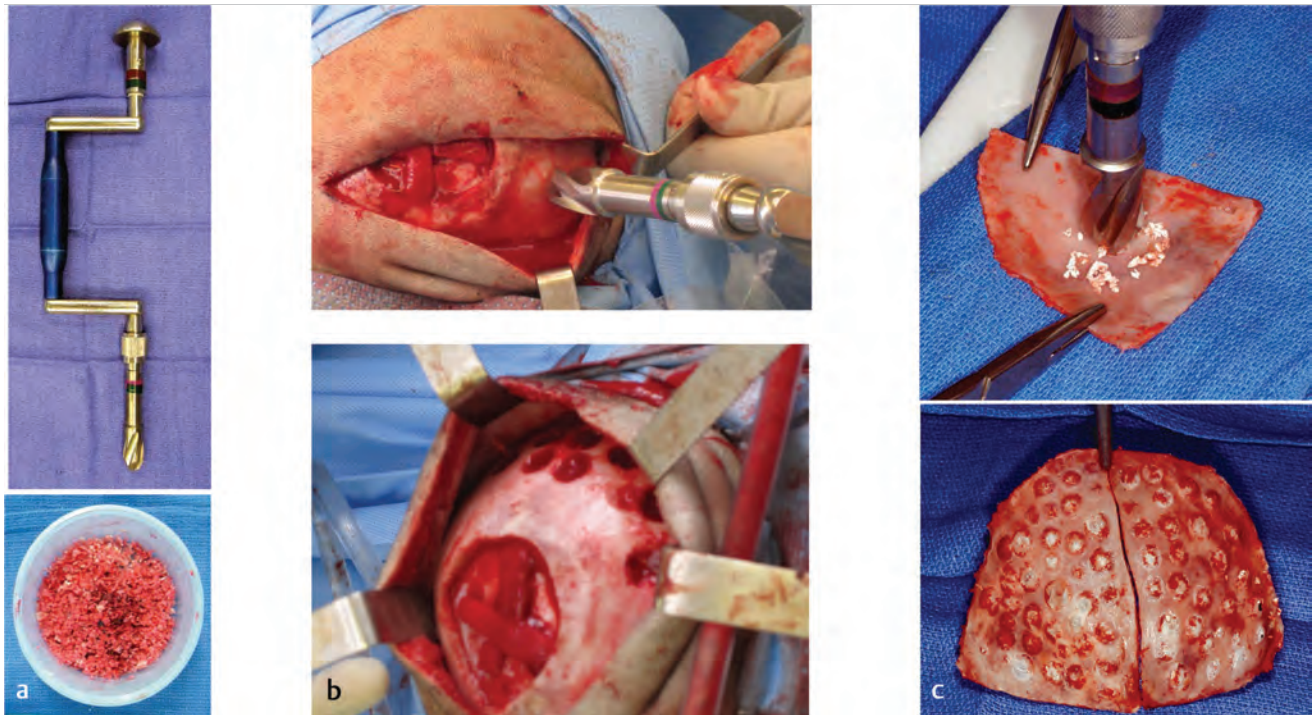


Fig. 8.3 Technique of particulate bone grafting. (a) Brace and bit with graft material. (b) Ectocortical harvest. Note partial-thickness donor-site defects. (c) Endocortical harvest.

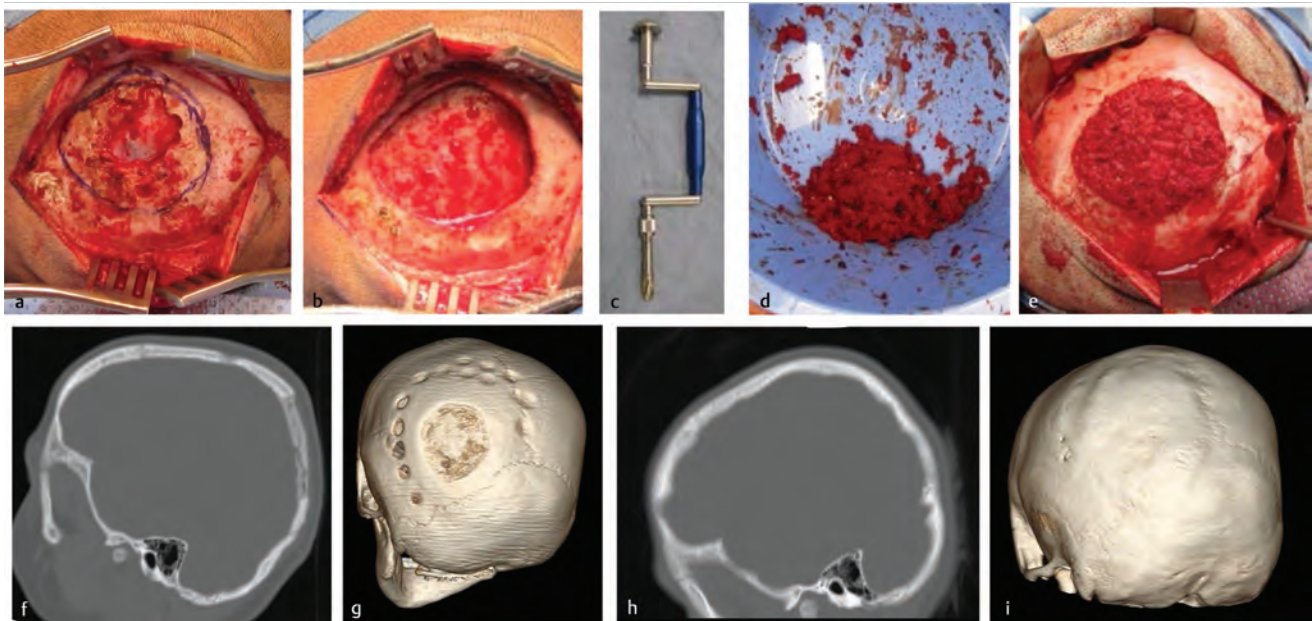


Fig. 8.4 Inlay cranioplasty using particulate bone graft in a 2-year-old child. (a) Venous malformation of the bone. (b) Defect following removal of the lesion. (c) Hudson's brace. (d) Particulate bone graft harvested from the ectocortical surface. (e) Graft placed over the dura. (f,g) CT images 6 weeks postoperatively. (h,i) Healed graft 2 years postoperatively. Note that the graft is indistinguishable from the adjacent cranium and completely osteointegrated, and the donor-site defects have healed.

thickness and less than critical size. Thus, they reossify and can be used again to harvest additional bone (reparative osteogenesis). Through cycles of harvest and healing, the craniofacial surgeon can access a nearly endless supply of autogenous cranial bone. Particulate graft is different from bone dust harvested with

a high-speed burr—this material produces much smaller particles that lack viable cells and are usually completely resorbed.

Disadvantages of particulate graft are that it does not provide immediate strength, is less amenable to correcting visible contour deformities of the frontal bone, and has a greater

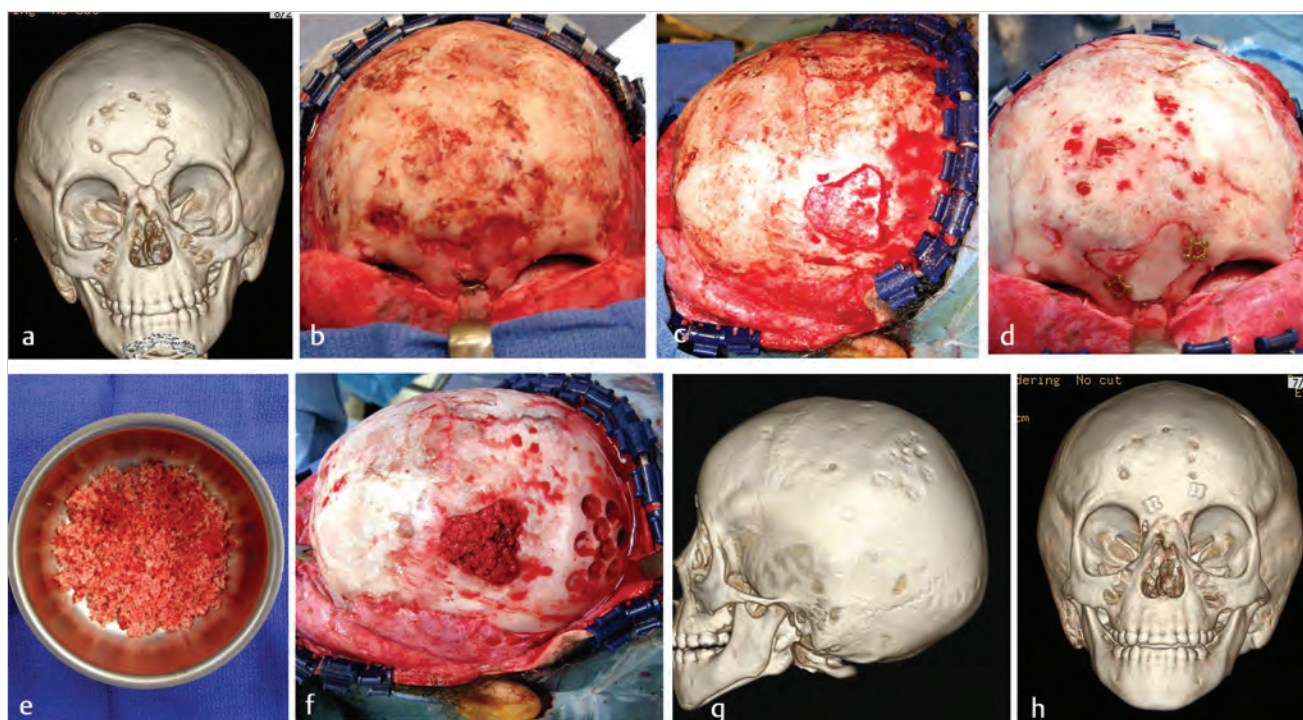


Fig. 8.5 Exchange cranioplasty in a 3-year-old child with a frontal bone defect following removal of a dermoid cyst complicated by an infection. Exchange cranioplasty was chosen instead of direct particulate bone grafting, because the dura at the frontal bone defect was scarred and the visible area of the forehead was a favorable location for a cortical graft. (a,b) Preoperative appearance of the defect. (c,d) Full-thickness parietal bone graft harvested and placed into frontal defect. (e,f) Particulate bone graft was harvested from the adjacent ectocortex and used to fill the parietal bone graft site. (g,h) Healed recipient and donor sites. (With permission from Greene et al 2008.)

resorption when placed over scarred dura. These limitations can be overcome by using exchange cranioplasty (► Fig. 8.5 and ► Fig. 8.6). This method involves harvesting a full-thickness piece of parietal bone and placing it over the defect. The donor site is then filled with particulate graft harvested from the adjacent ectocortex or endocortex of the parietal graft.

Almost any size defect can be managed with direct particulate bone grafting or exchange cranioplasty. An alternative to particulate bone/exchange cranioplasty in older children with a diploic space is split cranial bone grafting. A disadvantage of this technique is that the donor- and recipient-site bones are thinner than the surrounding cranium, and in older children, these sites cannot be used in the future to harvest additional bone graft. The thickness of split cranial bone can be augmented with particulate graft harvested from the ectocortical surface. The graft is placed under the cortical bone segment. Particulate graft offers some advantages over split cranial bone; for example, neurosurgical assistance is not required and particulate graft can also be harvested from those areas of the cranium that would be difficult to split (e.g., temporal, greater sphenoid wing, and occiput).

In our opinion, the use of nonautologous bone for inlay cranioplasty in children should be avoided because of the risk of dislodgement, infection, and growth restriction (► Fig. 8.7). The use of particulate graft has simplified the ability to perform autologous cranioplasty in the pediatric population. We do not advocate for the use of bone morphogenetic protein, because this osteoinduction agent may increase the risk of malignant transformation and craniosynostosis.

Adults

Many surgeons prefer autologous cranioplasty in adults. Unlike synthetic material, the healed bone graft has the same properties as native bone: strength, growth potential, stability, and resistance to infection. Split calvarial bone and/or particulate graft/exchange cranioplasty are our first-line techniques. However, because the cranium is no longer growing in adults, the use of nonautologous materials is much more prevalent compared with that in children. Alloplastic materials shorten operative time, obviate donor-site morbidity, and can be custom-made. These substances are particularly useful for patients with significant comorbidities.

The most commonly used alloplastic materials for inlay cranioplasty are titanium, hydroxyapatite, methylmethacrylate, and porous polyethylene. Titanium has a low risk of infection and is typically used for small defects, because it does not provide adequate strength to protect the brain. It also can be used as a temporary barrier to separate the brain from an overlying soft-tissue reconstruction (► Fig. 8.8). Hydroxyapatite, methylmethacrylate, and porous polyethylene are commonly employed but should not be placed in continuity with mucosae/sinuses, because they are at risk for infection. Hydroxyapatite also has a higher likelihood of fracture/displacement when used for large areas. Methylmethacrylate is usually not applied for inlay cranioplasty, because the heat generated from preparing the material can injure the brain. Medpore is our preferred nonautologous inlay cranioplasty material, because it can be easily contoured to the defect and allows fibrovascular ingrowth and some osseointegration.

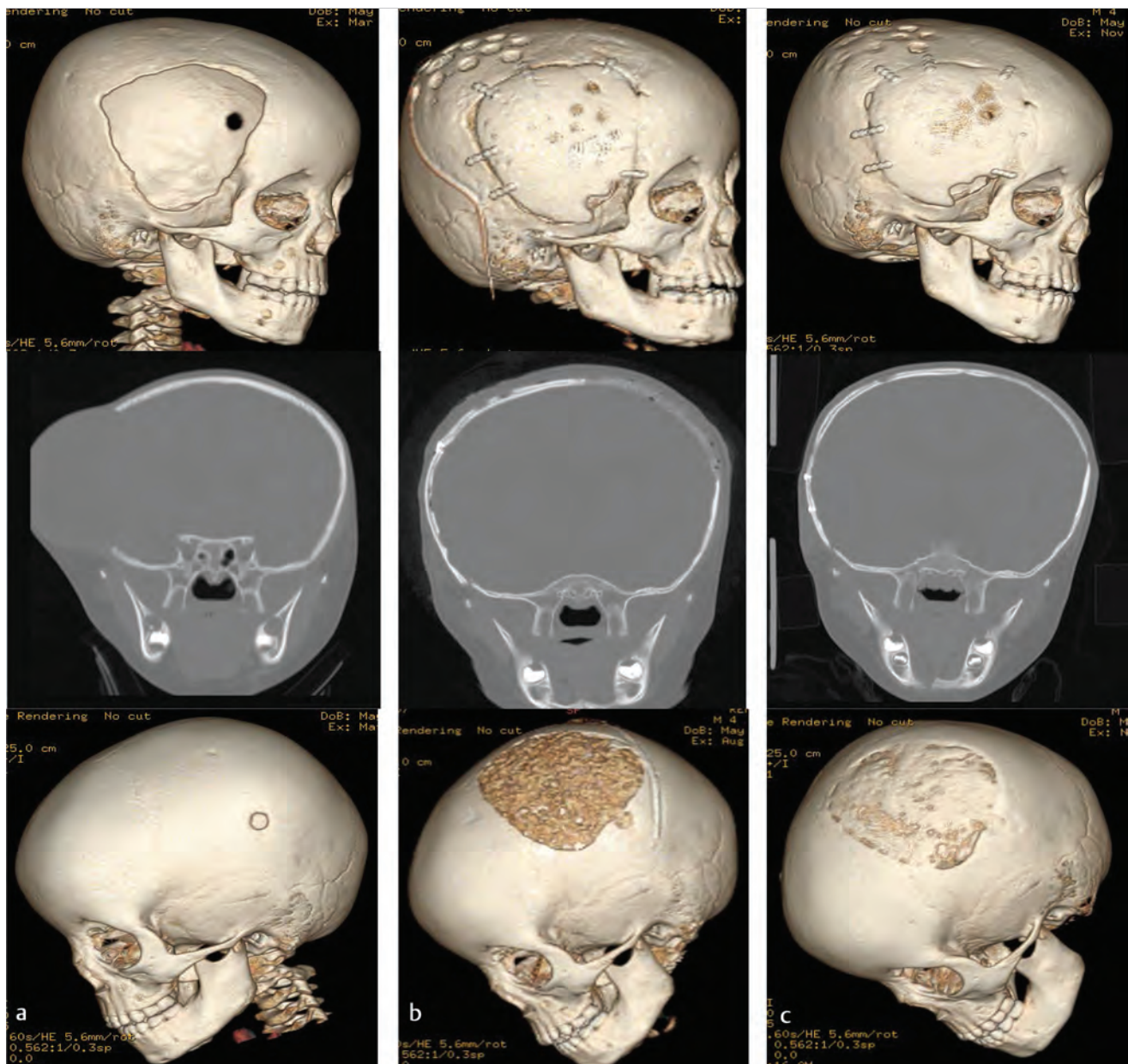


Fig. 8.6 A 4-year-old child with a large cranial defect following a motor vehicle accident. (a) Appearance before undergoing exchange cranioplasty. (b) Immediate postoperative CT images to rule out hydrocephalus and dural injury show full-thickness parietal graft over original defect and particulate graft in the parietal donor site. (c) Healed grafts 3 months postoperatively.

8.5 Complications

Although iliac and rib graft are rarely used for cranioplasty, donor-site problems can include neuropathic pain, pneumothorax, and deformity. When harvesting split cranial bone, the dura (and less commonly a subdural vein) can be injured. When a full-thickness piece of cranium is removed, a neurosurgeon should be present. Cranial bone grafts used for onlay cranioplasty have a high rate of resorption and may require additional augmentation procedures. Split cranial and particulate bone grafts used for inlay cranioplasty may not completely heal the defect and thus repeat grafting may be required. Infection of autologous bone grafts is rare; the risk of infection, displacement, and fracture of alloplastic substances is greater. If an

alloplastic material becomes infected, it cannot usually be salvaged with antibiotic therapy and requires removal, clearance of the infection, and then repeat cranioplasty.

8.6 Conclusion

The two most important variables to be considered for cranioplasty are (1) whether the problem is related to contour (onlay) or protection of the brain (inlay) and (2) the age of the patient (► Fig. 8.9). Because onlay cranioplasty is generally performed to improve an aesthetic deformity, we prefer to intervene when children are older and use an alloplastic material, because there is less resorption compared to bone grafts. We favor porous polyethylene, because it allows fibrovascular ingrowth and is

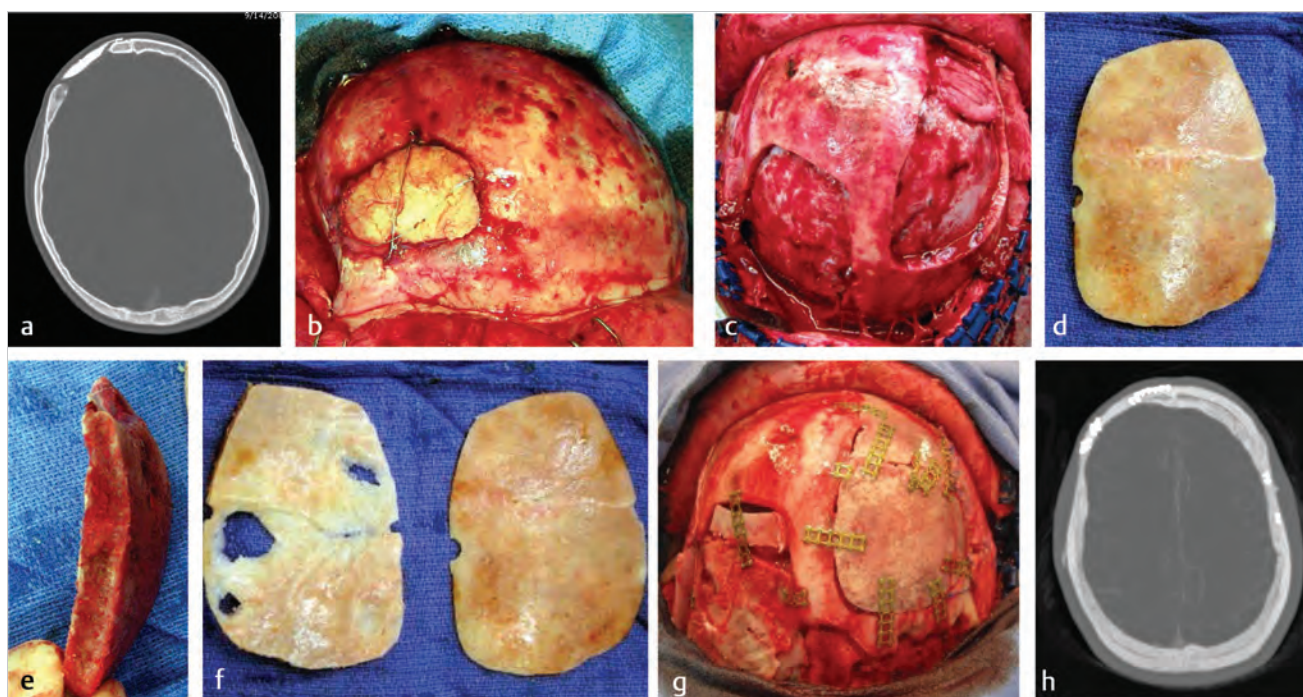


Fig. 8.7 A 9-year-old child managed with inlay methylmethacrylate at an outside hospital presented at the age of 13 years with pain and an unstable implant. (a,b) Preoperative image shows nonintegration of the implant with the adjacent cranium. (c–g). Patient was managed by removal of the alloplastic material and cranioplasty using split parietal bone graft. (h) Osteointegration of the bone graft postoperatively.

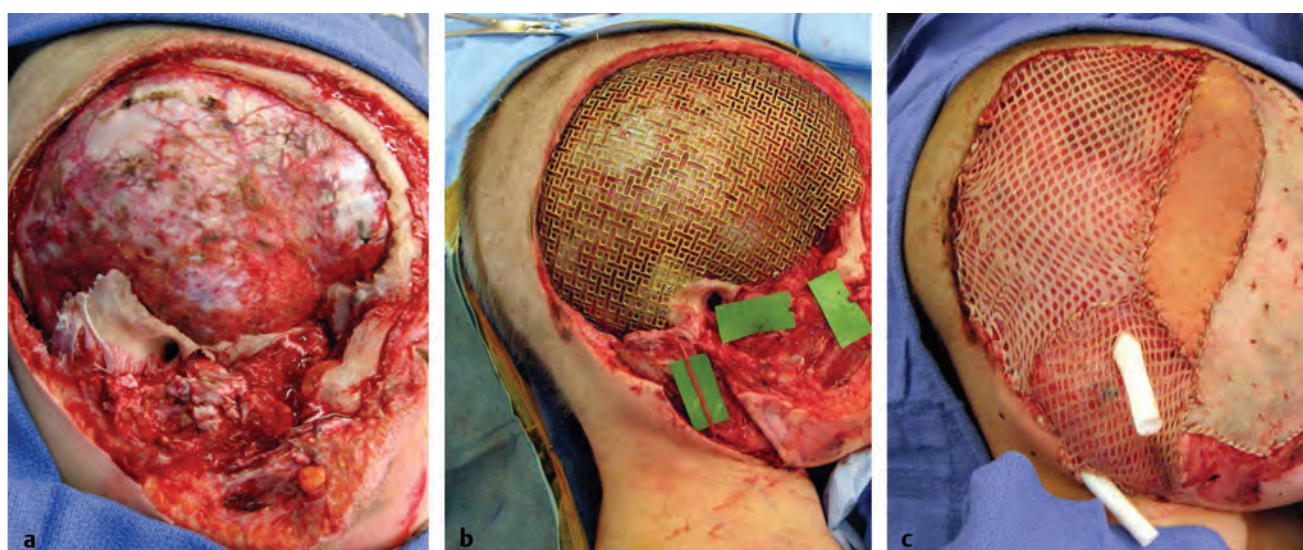


Fig. 8.8 Inlay cranioplasty using titanium. (a) A 5-year-old child who suffered a traumatic injury from a motor vehicle accident. (b,c) The brain was temporarily covered with titanium mesh to support a free flap to provide soft tissue coverage.

easy to contour. Large full-thickness cranial defects necessitate closure to protect the brain. We allow spontaneous ossification to occur during infancy and intervene after this time. Particulate bone grafting/exchange cranioplasty is our first-line technique in children and adults, because the bone grows with the child and completely osseointegrates. In adults with significant morbidities, we will consider inlay cranioplasty using alloplastic materials to limit the extent of the operation (► Table 8.1).

8.7 Key Points

- The cranium is the preferred autologous donor site for cranioplasty; there are three main types of grafts (full thickness, split thickness, particulate).
- Alloplastic materials are a good choice for onlay cranioplasty, especially in older children, because they undergo lesser resorption than bone grafts.

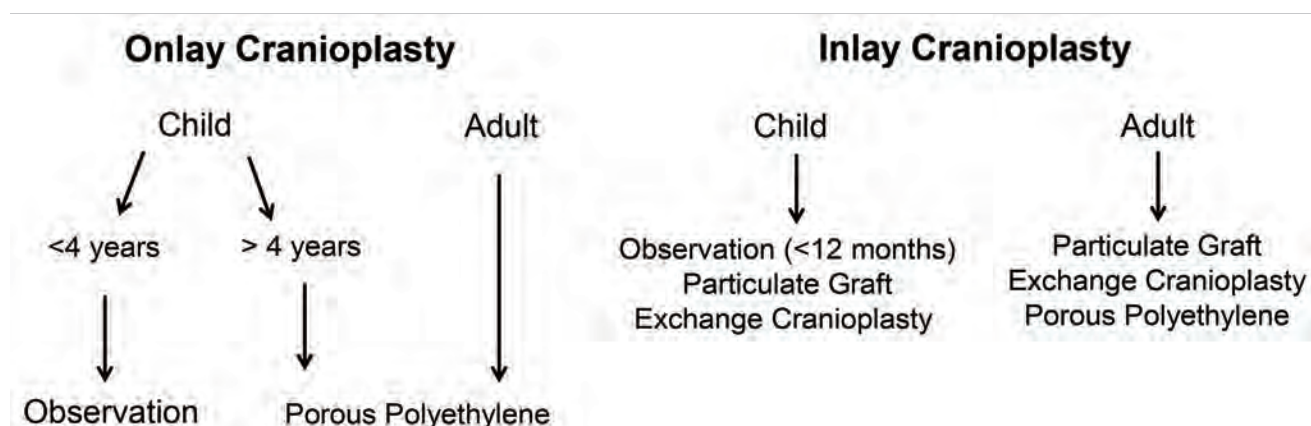


Fig. 8.9 Management algorithm for cranial defects.

Table 8.1 Comparison of authors' preferred materials used for cranioplasty

	Particulate graft	Split cranial graft	Porous polyethylene
Advantages	Does not require a diploic space for harvest Can harvest from any area of cranium Bone becomes as thick as surrounding cranium Donor and recipient sites can be reharvested Simpler than split cranial graft	Heals over scarred dura Immediate strength and contour	No donor-site morbidity Reduced operative time No resorption Peripheral fibrovascular ingrowth
Disadvantages	Less effective if placed over scarred dura No immediate strength/contour (Disadvantages are overcome by using exchange cranioplasty)	Requires diploic space for harvest Limited donor-site areas Not as thick as surrounding cranium	Does not completely osseointegrate Increased infection rate Cranial growth restriction and dislodgment if inlay in children

- Cranial bone grafts are better than alloplastic materials for inlay cranioplasty, because they osseointegrate, grow with the patient, and provide good strength and resistance to infection.
- Our preferred onlay cranioplasty material is porous polyethylene, because it allows fibrovascular ingrowth and is easy to use.
- Our inlay cranioplasty methods are particulate bone grafting/exchange cranioplasty and split cranial bone.

Suggested Readings

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9 Osseous Lesions of the Craniofacial Region

Salim Afshar

Summary

Osseous lesions in the craniofacial region can be discovered in a variety of clinical and radiological scenarios and often present as facial asymmetry or as a disturbance in growth such as tooth eruption. Having a broad understanding of the range of lesions that can occur in this area of the body is critical given that benign lesions can often be destructive and result in significant morbidity. Early diagnosis and a multidisciplinary team approach is often warranted in order to optimize results for the growing child.

Keywords: buccal, bifurcation, cyst, odontogenic, aneurysmal, nasopalatine, cementoblastoma, odontoma, cherubism, osteoblastoma, myxoma, neurofibroma

9.1 Introduction

Osseous lesions in the craniofacial region can be discovered in a variety of clinical and radiologic scenarios. Most commonly, when a pediatric patient is found to have a facial asymmetry or a mass, initial consultation is with a plastic surgeon or a maxillofacial surgeon. Having a broad understanding of the range of lesions that can occur in this area of the body is critical to understanding appropriate workup and surgical management. Because of the intimate association of lesions in this area with vital structures such as the jaw, sinuses, orbits, and ears, a multidisciplinary team approach is often warranted.

In this chapter, we classify osseous lesions of the craniofacial region broadly into cysts and tumors and further subdivide these lesions into odontogenic and nonodontogenic cysts and tumors.

9.2 Odontogenic Cysts

Most commonly, odontogenic cysts and lesions are discovered when there is radiographic investigation for asymmetric or delayed eruption of adult teeth. It is important to know the timing of eruption, and if there is significant delay between the right and left sides, then a radiologic examination is necessary. The most common finding is a cyst associated with an impacted

and displaced tooth. Odontogenic cysts are broadly divided into inflammatory and developmental cysts.

In the pediatric population, the most common developmental cyst is a dentigerous cyst associated with an impacted canine, a premolar, or a molar such as the third molar (► Fig. 9.1a). On a radiograph, there is a well-circumscribed, unilocular radiolucency associated with the crown of an unerupted tooth attached at the cemento-enamel junction (CEJ). The cyst can cause tooth displacement, and the treatment consists of enucleation of the cyst and, in most cases, removal of the impacted tooth (► Fig. 9.1b, c).

The odontogenic keratocyst and the adenomatoid odontogenic cyst were previously considered developmental cysts but are no more considered tumors. They are now known as keratocystic odontogenic tumor (KCOT) and adenomatoid odontogenic tumor (AOT) and will be discussed in the section on odontogenic tumors.

9.2.1 Inflammatory Cysts

Inflammatory cysts are often associated with a nonvital tooth and are painful on palpation and percussion. They can be asymptomatic if the lesion is chronic in nature and are usually associated with the roots of a nonvital tooth or a tooth with a large carious lesion. Treatment often involves root canal therapy of the tooth or extraction, with enucleation of the granulation tissue within the cyst.

9.2.2 Buccal Bifurcation Cyst

The buccal bifurcation cyst (BBC) is inflammatory rather than developmental and usually develops on the buccal region of vital mandibular first or second molars (► Fig. 9.2). It is usually seen in children between the age of 5 and 15 years. Treatment options include curettage of the lesion with daily irrigation or enucleation and removal of the tooth. They can become secondarily infected and may require antibiotics and surgical drainage of the surrounding tissue.

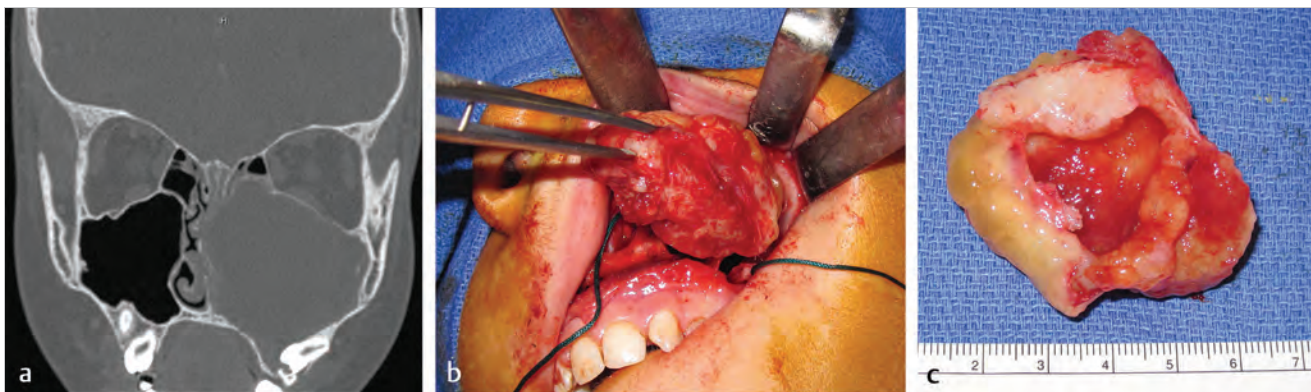


Fig. 9.1 (a) Large left maxillary cyst associated with an impacted premolar tooth. (b) Intraoperative view of lesion being removed from left maxillary sinus. (c) Specimen: Large left maxillary dentigerous cyst.

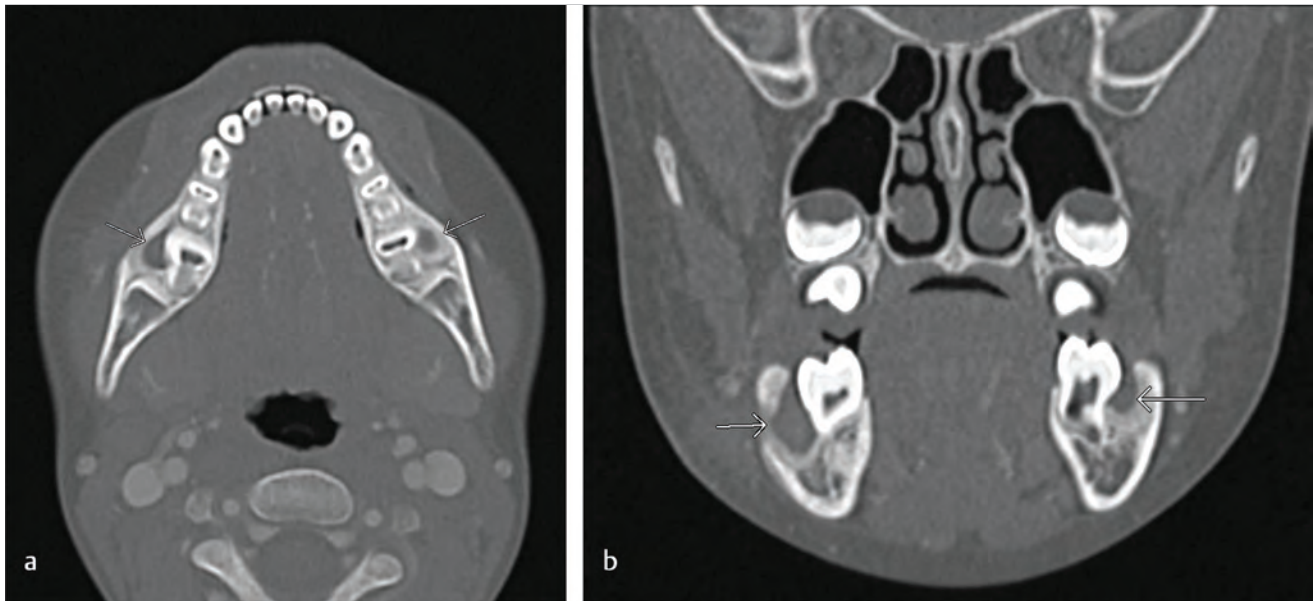


Fig. 9.2 (a,b) Axial view of bilateral buccal bifurcation cysts associated with erupting first molars.



Fig. 9.3 Simple bone cyst of the left mandibular ramus.

9.3 Nonodontogenic Cysts

The most common nonodontogenic cyst in childhood is the traumatic or simple bone cyst (SBC). Typically, the patient has an asymptomatic radiolucent lesion of the mandible, with no clinical swelling or any other abnormalities on examination (► Fig. 9.3). The lesion favors the male population and presents in the first two decades of life. The radiographic imaging warrants surgical exploration and curettage in order to rule out an epithelial lined cyst or tumor. The clinical finding is an empty bony cavity with very little tissue, if any, to submit to pathology. If the lesion is large enough or near the condylar head, a bone graft may be warranted, but typically, after curettage of the bony wall, the lesion will fill in with normal bone over time.

9.3.1 Aneurysmal Bone Cyst

Another similar radiographic presentation to a traumatic bone cyst is the aneurysmal bone cyst (ABC). This lesion tends to favor the female population, and often, there can be jaw swelling and pain. The most common type is the vascular type, which can be rapid growing and destructive (► Fig. 9.4a, b). Unlike a traumatic bone cyst, on entry of this lesion, you will encounter brisk bleeding, which is controllable with pressure and resolves with curettage of the lesion (► Fig. 9.4c–e). Aneurysmal bone cysts tend to have a recurrence rate of 10% and are part of a spectrum of vascular lesions; these cysts are associated with jaw tumors.

9.3.2 Nasopalatine Duct Cyst

Finally, the nasopalatine duct cyst (NPDC) is a nonodontogenic developmental cyst that is usually found incidentally in the anterior maxilla in the area of the incisive canal. It can become painful, especially if secondarily infected. Radiographically, on an occlusal plain film radiograph, one finds a midline heart-shaped unilocular radiolucency. The lesion often is symmetric and is present in the anterior maxilla along the length of the incisive canal (► Fig. 9.5). Treatment comprises enucleation and curettage, with recurrence being very rare.

9.4 Odontogenic Tumors

Odontogenic tumors are common in children because of the natural course of tooth development of the adult teeth, leading to eruption of the adult teeth and exfoliation of the deciduous teeth. As a result, the most common clinical signs of an odontogenic lesion are asymmetric tooth eruption and alveolar bone asymmetry.

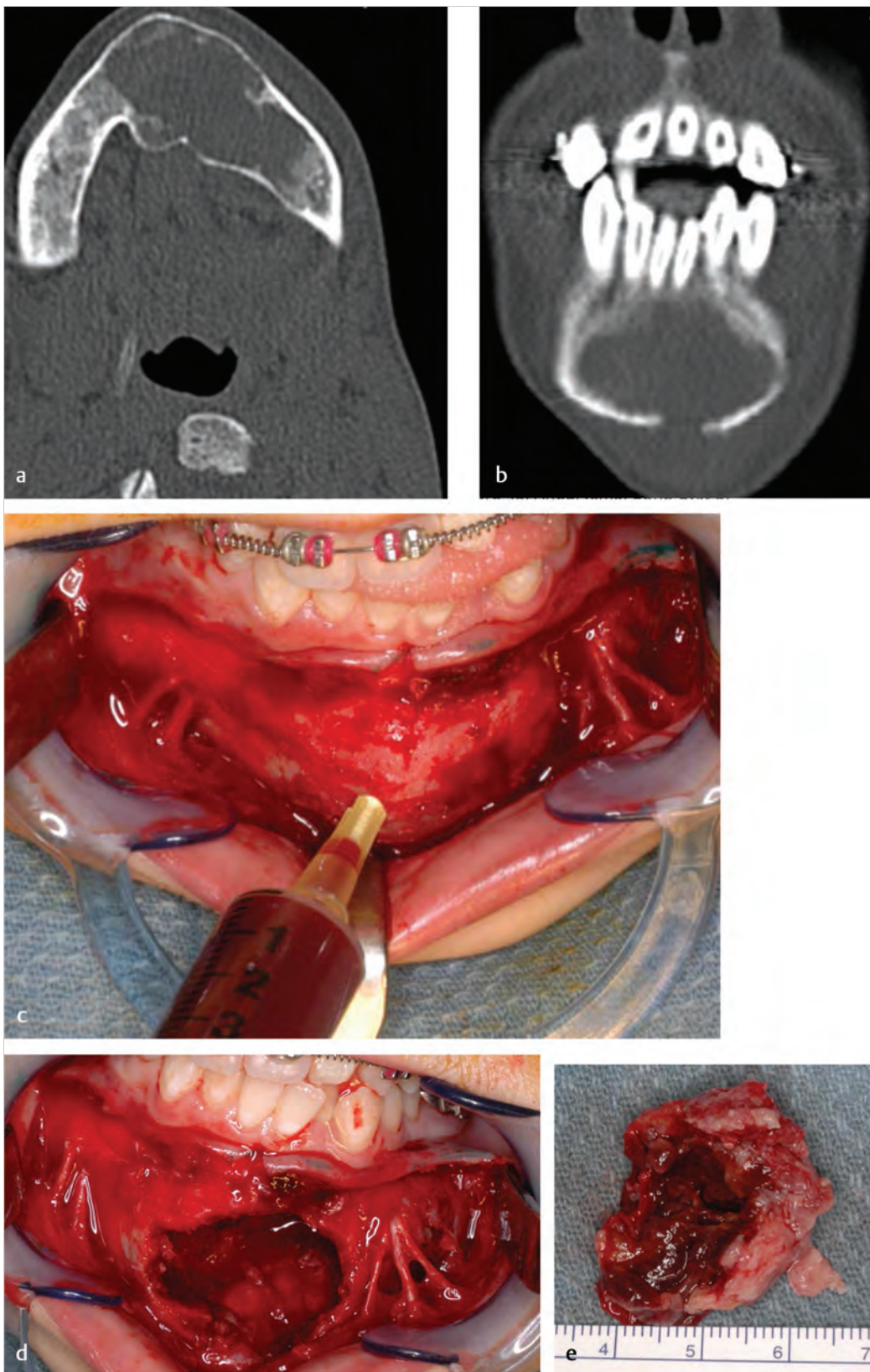


Fig. 9.4 (a) Aneurysmal bone cyst of the anterior mandible with bony expansion. (b) Coronal view of aneurysmal bone cyst of the anterior mandible. (c) Aspiration of aneurysmal bone cyst demonstrates vascular nature. (d) With removal of the lesion, brisk bleeding is resolved. (e) Specimen.



Fig. 9.5 (a) Nasopalatine duct cyst seen as an expansile lesion of the incisive canal of the maxilla. (b) Coronal view of lesion.



Fig. 9.6 (a) Odontoma of the maxilla adjacent to an erupting canine and lateral incisor. (b) Specimen: Odontoma of the maxilla.

9.4.1 Odontoma

The most common odontogenic tumor of the jaws is the odontoma. There are two types of odontomas: compound and complex. The compound odontoma consists of multiple toothlike structures surrounded by a radiolucent zone, whereas the complex odontoma is a radiodense mass made up of enamel and dentin without toothlike structures and is surrounded by a radiolucent zone. The mean age of people found with odontoma is 15 years, and odontomas are often asymptomatic and found on routine dental radiographs. They may cause delayed

eruption of teeth. Clinically, they are typically well-circumscribed dense masses that can be treated with simple surgical excision that must also include complete removal of associated soft tissue, follicular tissue, or cyst. Prognosis is excellent, without recurrence (► Fig. 9.6).

9.4.2 Cementoblastoma

Similar to odontomas in appearance, cementoblastoma tends to appear radiographically in patients younger than 20 years as a round radiopaque mass fused to the roots of vital teeth

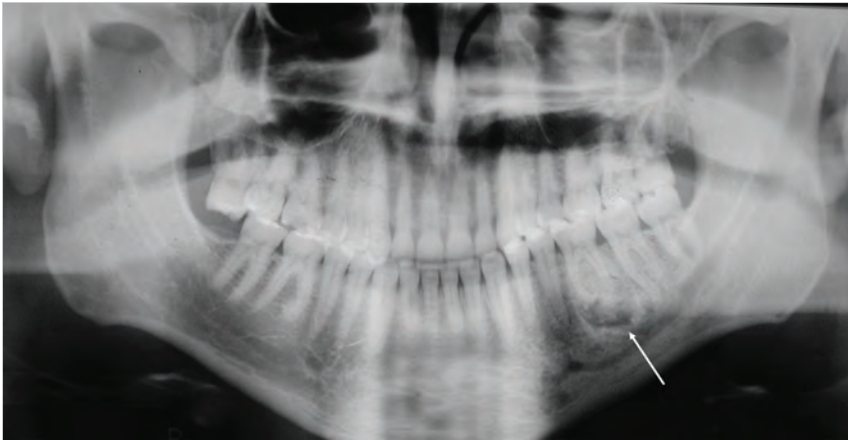


Fig. 9.7 Cementoblastoma associated with root of left mandibular first molar.

(► Fig. 9.7). This is a rare benign tumor of odontogenic origin and is treated with surgical excision of the mass and the affected tooth or teeth, followed by curettage or peripheral ostectomy. Cementoblastomas tend to recur if not completely removed, with reported recurrence rates near 40%.

9.4.3 Keratocystic Odontogenic Tumor

The keratocystic odontogenic tumor is a benign intraosseous cystic tumor of odontogenic origin, and it can be locally destructive. Majority of the lesions occur in the posterior mandible, with bony expansion and tooth displacement. Depending on the size and location, small lesion can be treated with enucleation and peripheral ostectomy, whereas larger KCOTs should be first decompressed, enabling the placement of a stent, followed by daily irrigation for 9 months, which reduces the lesion's size and causes dedifferentiation of the residual epithelium, thus enabling cystectomy. It has lower recurrence rates of around 10% (► Fig. 9.8). This is useful in children with multiple lesions and/or lesions that are in close proximity to vital structures such as the inferior alveolar nerve. Finally, marginal or en bloc resection can be utilized for large, recurrent lesions. Recurrence rates range between 10 and 55%, with enucleation alone having the highest rate.

9.4.4 Nevroid Basal Cell Carcinoma Syndrome

When multiple KCOTs are present, the child must be further worked up for nevoid basal cell carcinoma syndrome (NBCCS), also known as Gorlin–Goltz syndrome. Keratocystic odontogenic tumors occur in 75% of patients with NBCCS. Keratocystic odontogenic tumors that occur as a part of this syndrome tend to have a higher recurrence rate.

9.4.5 Adenomatoid Odontogenic Tumor

The adenomatoid odontogenic tumor is a benign cystic hamartoma that is known as the “two-thirds” tumor, because about two thirds occur in maxilla, two thirds occur in young women (preteenagers and teenagers), two thirds are associated with impacted tooth, and two thirds of those teeth are canines.

They can become very large and radiographically appear as a well-demarcated, unilocular radiolucent lesion usually associated with an impacted tooth. Treatment involves direct exploration and enucleation, ensuring that the capsule is intact upon removal. Recurrence is rare.

9.4.6 Ameloblastic Fibro-odontoma

Ameloblastic fibro-odontoma (AFO) is usually seen in teenagers as an asymptomatic jaw expansion that may resorb adjacent teeth. It has a mixed radiolucent–radiopaque presentation and is radiographically similar to a developing odontoma (► Fig. 9.9a). Majority of these tumors are found in the posterior mandible and are often associated with an unerupted tooth. Treatment includes simple enucleation and curettage (► Fig. 9.9b, c). Prognosis is excellent, and recurrence is rare.

9.4.7 Ameloblastic Fibroma

Ameloblastic fibroma (AF) is usually seen between 6 and 15 years of age and usually presents as an asymptomatic expansion of the jaw in the molar region of the mandible. It can be of unilocular or multilocular radiolucency, occurs most commonly in the mandible, and is associated with an impacted tooth (► Fig. 9.10). Because it may have a similar appearance to other large destructive lesions, an incisional biopsy should be performed in children as an AF is treated with simple enucleation and curettage versus a resection for other lesions of similar appearance. Although rare, in some cases, an AF can convert into an ameloblastic fibrosarcoma. There is a significant higher risk of malignant transformation and recurrence in patients older than 20 years. As a result, resection is the treatment of choice in patients older than 20 years, patients with massive tumors, and those who have recurrent ameloblastic fibroma.

Ameloblastic fibrosarcoma is the most common malignant odontogenic tumor and is capable of metastasis. It tends to appear later than ameloblastic fibroma, usually in the 20s. Treatment involves resection by using 1.0 to 1.5 cm bony margins and one uninvolved anatomic carrier.

Ameloblastoma is the most common odontogenic tumor, with about 50% of them occurring between 20 and 40 years of age. In the pediatric population, ameloblastoma must be



Fig. 9.8 (a) Large left mandibular KCOT associated with impacted third molar. (b,c) Stenting of left mandibular KCOT.

considered in the differential diagnosis of any unilocular or multilocular radiolucency. Unicystic ameloblastoma represents about 5% of all ameloblastomas and tends to occur in younger age range (10–24 years) (► Fig. 9.11a, b). For all ameloblastomas, 80% occur in the mandible and 75% in molar–ramus region. They are benign, locally aggressive, expansile neoplasms that commonly cause dental changes such as mobility, tooth displacement, and root resorption. Untreated tumors can progress to a tremendous size. Treatment involves resection with 1.0 to 1.5 cm bony margins and one uninvolved anatomical barrier (► Fig. 9.11c, d). Consider frozen sections of soft tissue margins as well as taking a radiograph of the specimen to ensure that you have 1-cm margins past radiographic margins (► Fig. 9.11e). Cure rate with resection is about 98%. Controversy exists around immediate reconstruction at the time of resection versus a staged approach. If enucleation and curettage are performed, there is a recurrence rate of more than 80% (► Fig. 9.12).

9.5 Nonodontogenic Tumors

9.5.1 Giant Cell Lesions

Giant cell lesions in children and young adults usually present clinically as painless expansion of the anterior jaw, favoring women over men and the mandible over the maxilla. They present more often in the anterior mandible, with the canine–premolar area being the most common site (► Fig. 9.13). Giant cell lesions are classified as either nonaggressive or aggressive, based on criteria from clinical behavior and radiographic characteristics. It is critical to make the correct diagnosis as the treatment, and recurrence rate is substantially different between aggressive and nonaggressive lesions. For example, a nonaggressive giant cell lesion can be treated successfully with simple enucleation and curettage, which have low recurrence rates, whereas if the lesion were aggressive, the same treatment would result in a recurrence rate as high as 75%.

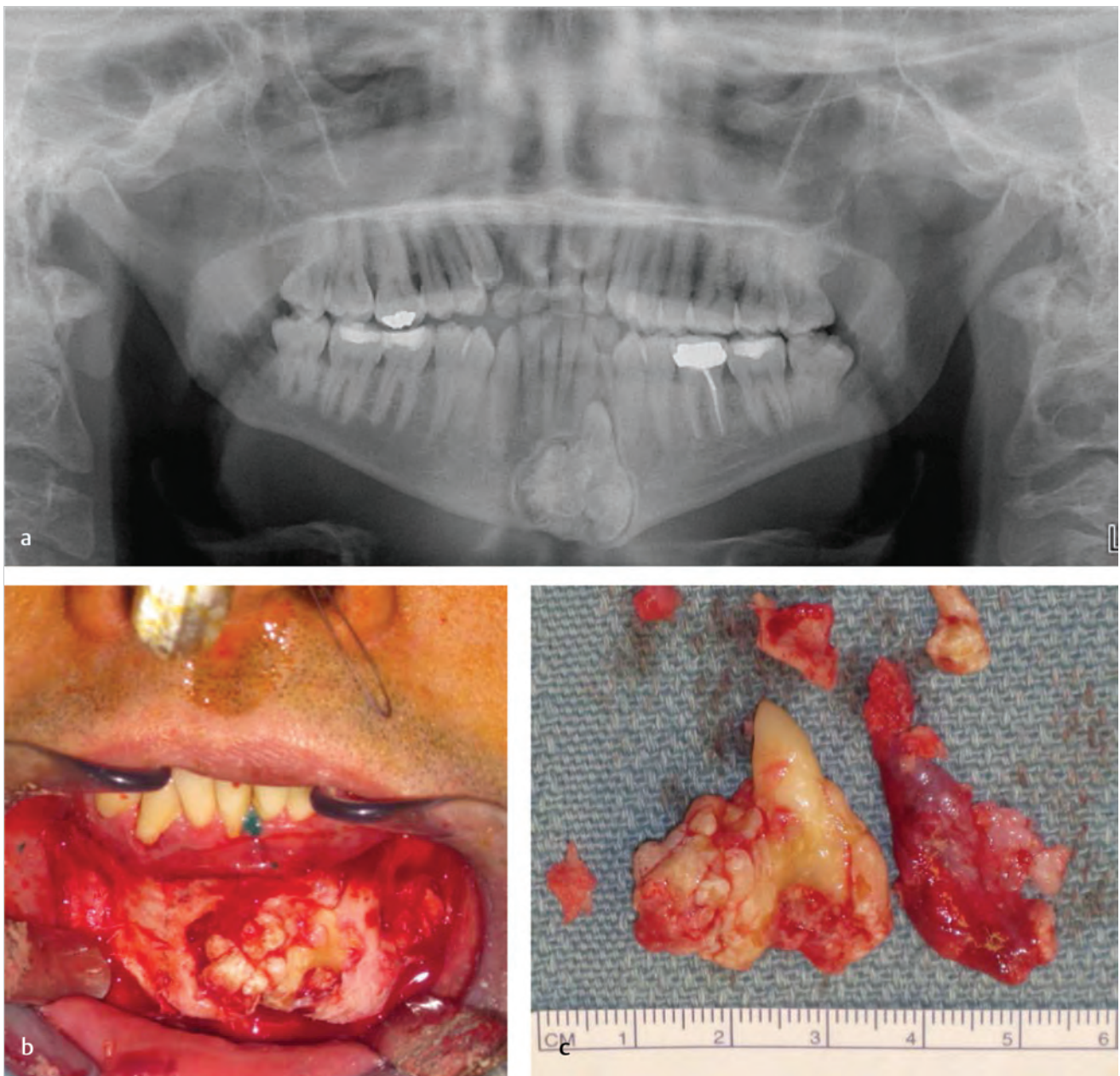


Fig. 9.9 (a) Ameloblastic fibro-odontoma of the anterior mandible, radiographically similar to an odontoma. (b) Intraoperative view of ameloblastic fibro-odontoma of the anterior mandible. (c) Ameloblastic fibro-odontoma after being removed.

Aggressive giant cell lesions tend to have a high recurrence rate and tend to occur in younger patients (mean age, 6 years). In order to be classified as aggressive, the lesion must be larger than 5 cm, be recurrent, or show three of the following six clinical characteristics: (1) rapid growth, (2) tooth loosening, (3) tooth displacement, (4) root resorption, (5) cortical thinning, and (6) cortical perforation. Treatment options vary from intralesional steroid injections and calcitonin injections to treatment with interferon- α . For a biopsy-confirmed giant cell lesion meeting the aggressive classification, the author recommends enucleation and curettage of the tumor and use of interferon- α -2 (3,000,000 units/m²), starting 48 to 72 hours postoperatively. Patients are then followed by clinical examination and radiography every 3 months, until bony cavity is completely

healed, and then after every 6 months. It is important to note that patients may develop infections, flulike symptoms, and other side effects of interferon therapy, requiring close monitoring by an experienced team. In patients in whom adjuvant therapy is not advisable, en bloc resection with 1.0 to 1.5 cm margins and one anatomic barrier is appropriate.

Nonaggressive giant cell lesions, when compared with aggressive lesions, tend to occur in slightly older children (mean age, 9 years) and are smaller in size (less than 5 cm). Lesions meeting the nonaggressive classification are appropriately treated with enucleation and curettage, with close monitoring to ensure bony fill. Bone grafts into the cavities are not typically recommended as the intact periosteum facilitates regeneration in young patients.

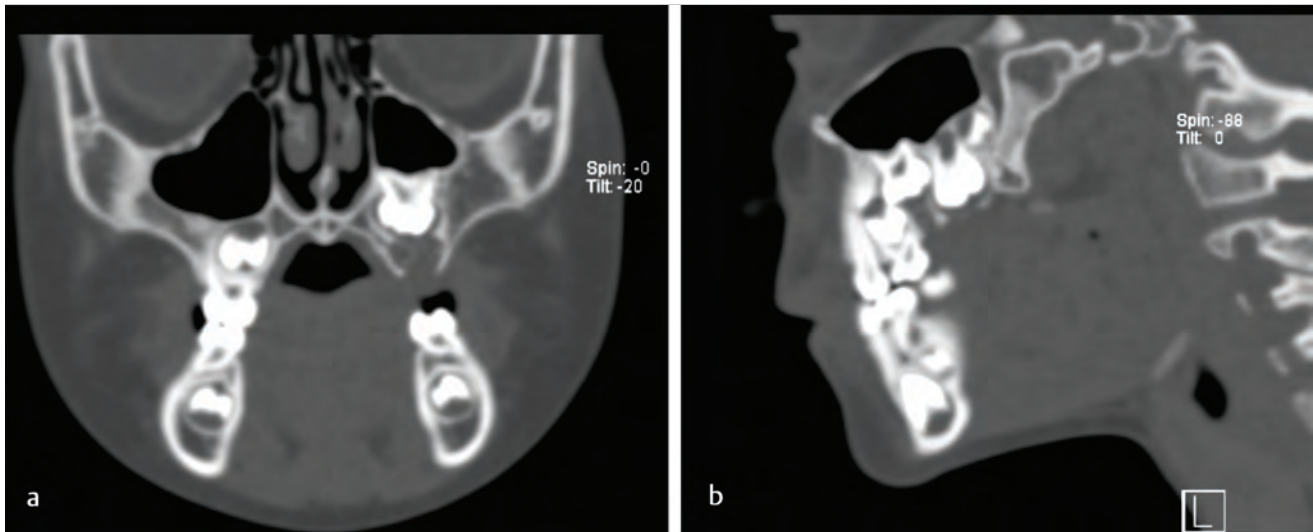


Fig. 9.10 (a) Ameloblastic fibroma of left maxilla associated with impacted first molar. (b) Ameloblastic fibroma—radiolucent lesion associated with impacted left maxillary first molar.

9.5.2 Brown Tumor of Hyperparathyroidism

The brown tumor of hyperparathyroidism is microscopically identical to the giant cell lesions described previously and therefore needs to be considered and ruled out. Lesions of this type are seen in patients with renal failure or secondary to hyperparathyroidism. Laboratory tests that should be ordered are serum calcium, parathyroid hormone (PTH), and alkaline phosphate. These lesions resolve once the underlying pathology is cherubism.

9.5.3 Cherubism

Patients with cherubism also have similar microscopically identical lesion to giant cell lesions. Cherubism needs to be considered if multiple giant cell lesions are found in a child (► Fig. 9.14). Cherubism favors males population over female population, presents early in life (2–5 years) and, in mild cases, can present later (10 years), and typically presents as bilateral painless expansion of the posterior mandible. The giant cell lesions often only involve the angle and ascending ramus of the mandible but not the condyle. Prognosis is variable, making treatment difficult. In some patients, early intervention with enucleation and curettage of the lesions results in excellent results, whereas in other patients, it is followed by rapid regrowth of the lesions and worsening deformity. Although cherubism is self-limiting by the age of 30 years, observation alone can occasionally result in severe facial deformity, with functional limitation and associated psychologic problems.

9.5.4 Multiple Giant Cell Lesion Syndrome

There is a subset of patients with Noonan's syndrome who also have multiple giant cell lesions, also referred as Noonan-like/multiple giant cell lesion syndrome (MS/MGCLS). These

patients often have short stature, wide forehead, ocular hypertelorism, prominent posteriorly angulated ears, high arched palate, webbed neck, and multiple giant cell lesions of the bones. Lesions associated with Noonan's syndrome may resolve after puberty, and therefore, conservative therapy is typically recommended.

9.5.5 Fibrous–Osseous Lesions

Fibrous–osseous lesions are a group of lesions that are known to affect the jaws and the craniofacial bones. This group of lesions shares a common process, in which the normal architecture of bone is replaced by fibrous tissue containing varying amount of mineralized tissue. Some of these lesions are felt to be neoplastic, whereas others are related to metabolic disturbances. They share common characteristic clinical, radiographic, and microscopic features. They are typically found in patients between 10 and 20 years of age and radiographically present as ovoid expansions of the jaw with ground-glass or orange-peel radiopaque appearance. Unfortunately, these lesions have been under frequent renaming and reclassification due to their varied features. In this chapter, we utilize the Speight and Carlos's classification from 2006 that is focused on histopathologic features. The main groups in this classification are the following: fibrous dysplasia (FD), osseous dysplasia (OD), and ossifying fibroma (OF).

9.5.6 Monostotic Fibrous Dysplasia

Fibrous dysplasia can be further classified as monostotic FD, craniofacial FD, and polyostotic FD. Monostotic FD accounts for about 70% of all lesions and involves only one bone (► Fig. 9.15). When lesions are in the maxilla and involve adjacent structures such as the zygoma and sphenoid, the designation craniofacial FD is utilized (► Fig. 9.16). When craniofacial bones are involved, cranial nerve compression can occur. Craniofacial FD tends to be in younger patients when compared with monostotic FD. Both typically present as a painless swelling that

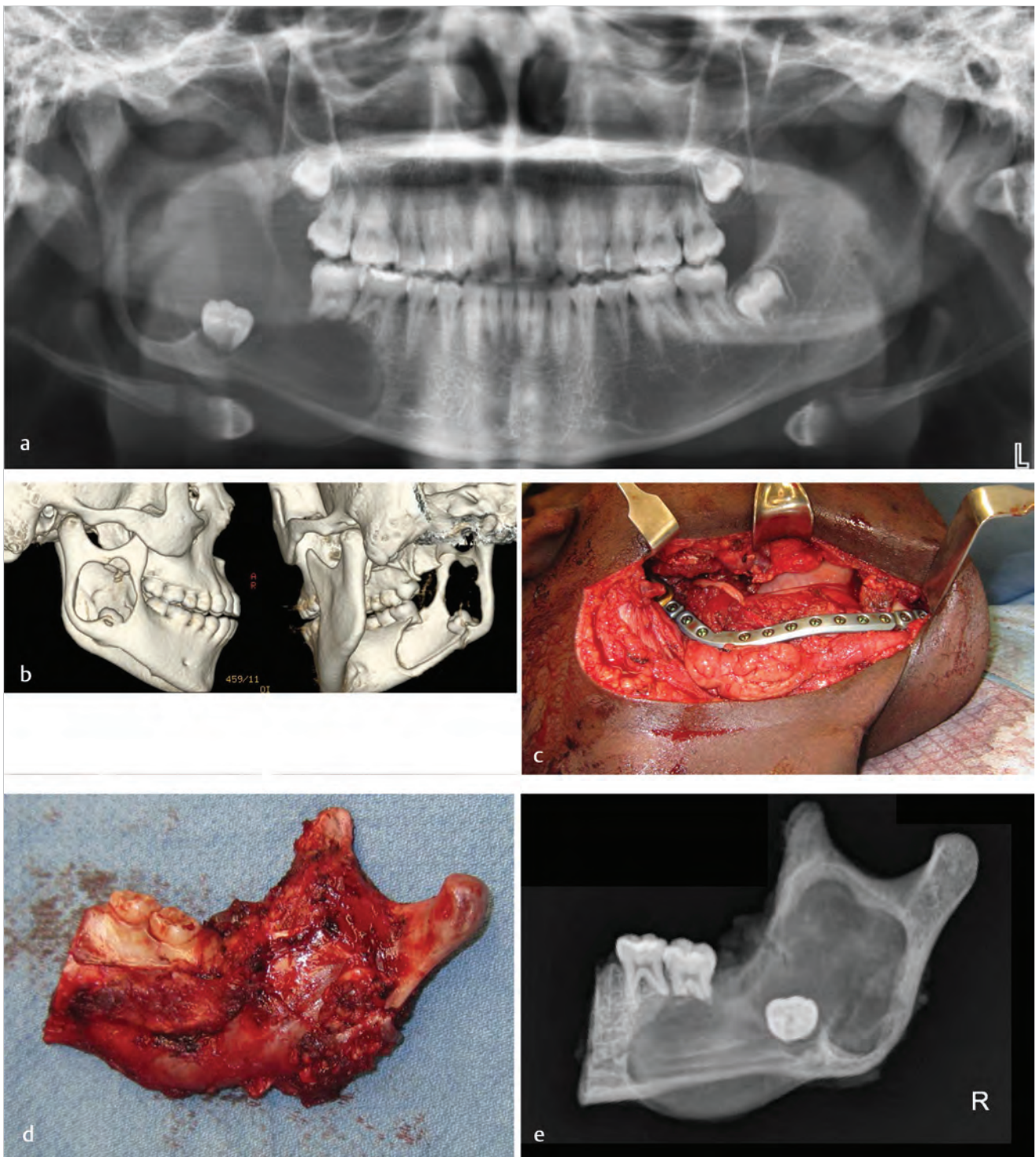


Fig. 9.11 (a) Unicyclic ameloblastoma of the right mandible. (b) Three-dimensional rendering of right mandibular ameloblastoma. (c) Resection of the right mandibular ameloblastoma and placement of a reconstruction plate. (d) Specimen figure. (e) Radiograph of specimen ensuring 1 cm bony margins.

demonstrates very slow growth. Teeth involved in the lesions are usually stable but may be displaced by the mass. Radiographically, FD is seen as a fine-ground-glass opacification that is not well demarcated from the surrounding bone and often involves the cortex (► Fig. 9.15). The final subtype of FD is polyostotic FD, which is characterized by involvement of two or

more bones. Two thirds of patients with polyostotic FD are younger than 10 years, and when seen with café au lait pigmentation, the condition is termed Jaffe–Lichtenstein syndrome. In addition, about 3% of patients with polyostotic FD have multiple endocrinopathies, a pattern known as McCune–Albright syndrome. All patients with polyostotic FD have symptoms that

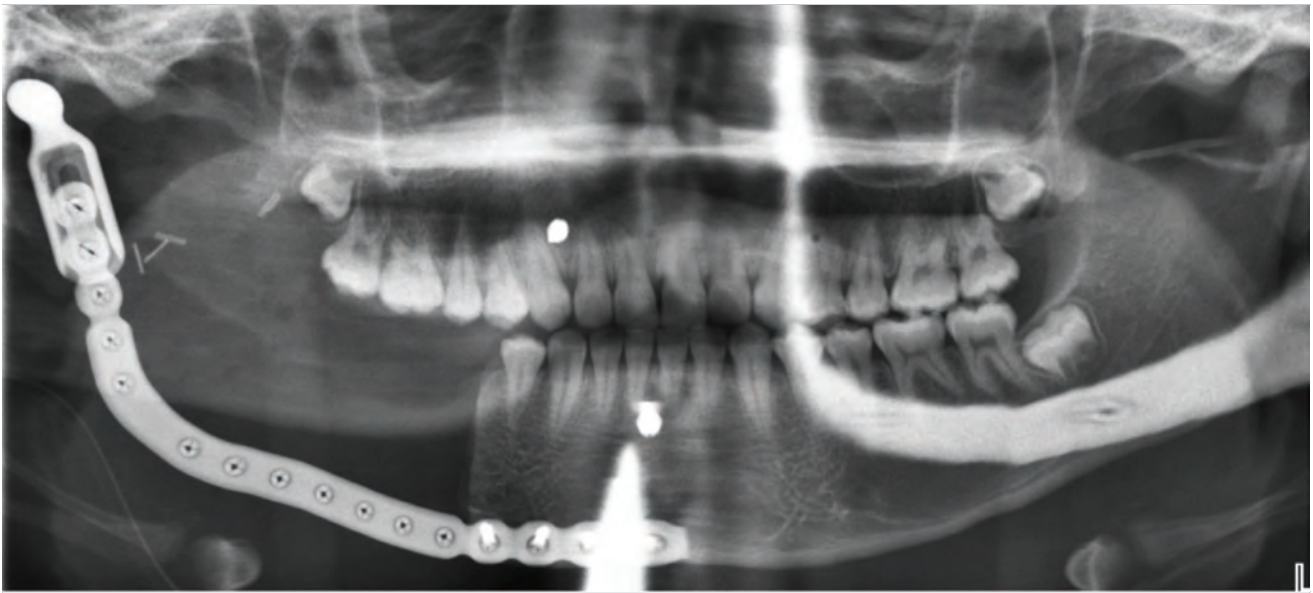


Fig. 9.12 Postresection radiograph.



Fig. 9.13 Three-dimensional rendering of CT demonstrating giant cell of anterior mandible.

tend to be dominated by the involved long bones and not the craniofacial region.

Overall, the treatment of FD varies depending on the clinical presentation, degree of deformity, and age of the patient. Smaller lesions, particularly in the mandible, may be surgically resected, but when lesions involve the zygoma, maxilla, and/or sphenoid, resection is not often possible and surgical reduction is the only option. Regrowth of the lesion often occurs over time

and is more common in younger patients. In rare cases, malignant transformation to osteosarcomas has been described, and they are more commonly associated with lesions that have received radiation therapy. Finally, bisphosphonates have been used in the treatment of FD with varied success, most consistently improving pain associated with FD.

9.5.7 Osseous Dysplasia

Osseous dysplasia is a group of fibrous–osseous lesions that includes periapical OD, focal OD, florid OD, and familial gigantiform cementoma. The first three, periapical, focal, and florid, do not present in the pediatric population and occur only in the tooth-bearing areas of the jaws. Familial gigantiform cementoma presents during the first decade of life as multifocal radiolucencies that eventually evolve into a mixed radiolucent, radiopaque pattern, ultimately resulting in massive sclerotic masses involving both the maxilla and the mandible. The masses tend to grow rapidly during adolescence, before entering the final slow sclerotic stage. Because of the rapid regrowth of the dysplastic tissue when in the mixed stage, treatments such as shave down and resection should not be done before the final sclerotic stage, when the lesions are predominately radiopaque.

9.5.8 Ossifying Fibroma

Ossifying fibroma includes conventional OF, juvenile trabecular OF, and juvenile psammomatoid OF. Conventional OF is a true neoplasm seen in adults, mainly in the premolar and molar areas of the jaw. In the pediatric population, juvenile OFs typically present before the age of 15 years, and both patterns have similar radiographic features and growth patterns. They tend to occur more commonly in the maxilla and often appear as a well-circumscribed, radiolucent lesion. Both types demonstrate slow growth, and the psammomatoid variant is usually outside



Fig. 9.14 (a) Multiple lesions—Noonan's syndrome. (b–d) Three-dimensional rendering of multiple lesions—Noonan's syndrome.



Fig. 9.15 (a,b) Fibrous dysplasia.

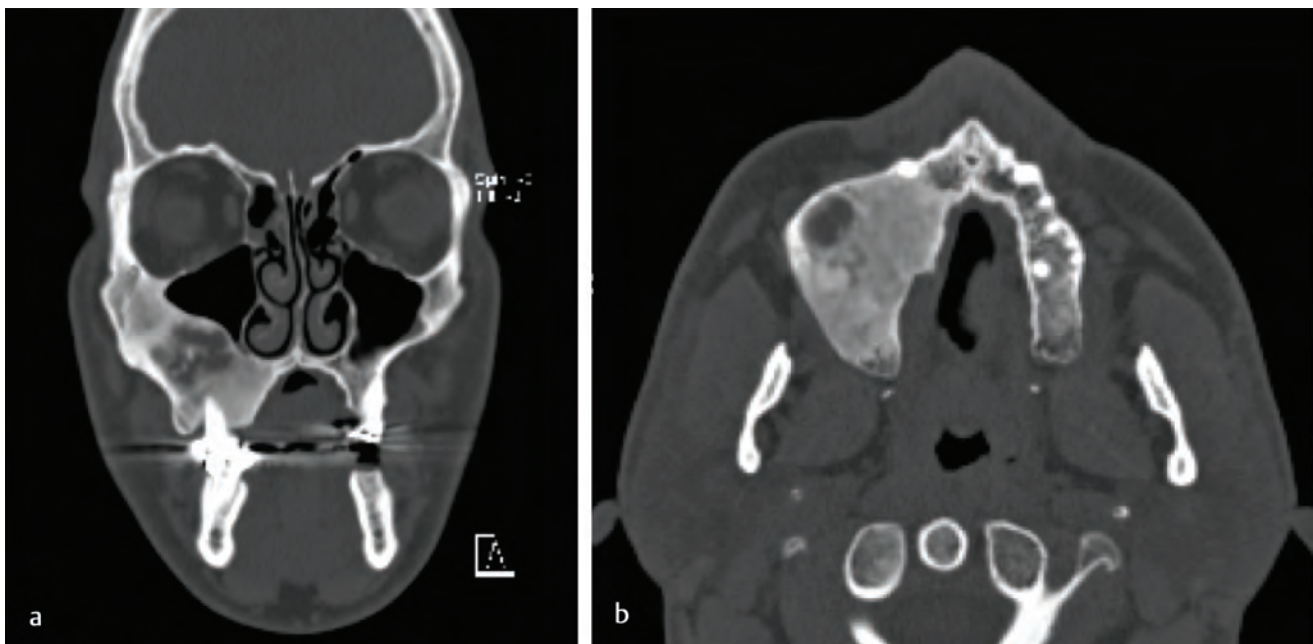


Fig. 9.16 (a,b) Craniofacial fibrous dysplasia.

of the jaws, with more than 70% occurring in the orbital, frontal, and paranasal sinuses. Typically, complications from this lesion are secondary to impingement of adjacent structures, resulting in nasal obstruction, exophthalmos, or proptosis. Clinical management of juvenile ossifying fibroma is unclear, with some lesion demonstrating rapid aggressive growth, especially in infants and young children. Small lesions may be locally excised, whereas larger lesions may require resection. Unlike the adult conventional form, juvenile OFs tend to have a higher recurrence rate, and close follow-up is required.

9.5.9 Osteoblastoma

Osteoblastoma occurs rarely in the jaws. It is more common in the vertebrae and long bones and tends to present in children and young adults. Patients typically experience swelling and have a history of chronic dull pain. The calvarium is the second most common site, and patients who have the temporal bone or zygoma involved may have secondary trismus. Clinically, patients present with bony expansion, mild, dull pain that is tender to palpation, and, if the mandible is involved, possible



Fig. 9.17 Osteoblastoma.

paresthesia. Radiographically, they present as mixed radiolucent–radiopaque lesions (► Fig. 9.17). Recommended treatment is wide en bloc resection with 1.0 cm bony margins, including one anatomic soft tissue layer, when possible. Lesions may slowly recur up to 10 years after resection. With enucleation and curettage alone, there is a high recurrence rate of about 25%.

9.5.10 Myxoma

Myxoma present as a rapidly growing, painful jaw mass, most commonly in the mandibular angle and ramus, in children. It has a clinical and radiologic presentation similar to ameloblastoma. Peak incidence is between 15 and 25 years of age, but unlike ameloblastoma, this tumor frequently presents in children younger than 10 years. The rapid growth and pain in children often result in a misdiagnosis of an acute infection. Radiographically, myxoma is present as a multiloculated radiolucency with soap-bubble appearance, typically within the posterior jaw. Cortical perforation and soft tissue involvement are often present. Treatment involves 1.0 to 1.5 cm margins and one uninvolved anatomic barrier, but others have advocated

smaller margins in pediatric population due to the resulting facial disfigurement and disruption in facial growth. Recurrence is as high as 25% and typically within the first 2 years after treatment. Myxomas can also occur in other areas of the face, such as the paranasal sinus. Midfacial myxomas are rare and should be differentiated from other benign and malignant tumors such as dermoid and rhabdomyosarcomas.

9.5.11 Aggressive Fibromatosis

Aggressive fibromatosis, also known as a desmoid tumor when located within the abdomen, is a benign but locally aggressive tumor found in children. Diagnosis is usually between 8 and 15 years of age, and it often presents in the head and neck region. The mandible is most commonly affected, and radiographically, it presents as a radiolucent, destructive lesion causing erosion of the adjacent cortices. Clinically, it often presents as a rapidly growing, painless mass in the head and neck region. Because of its local infiltration, treatment requires en bloc resection. Recurrence rate is up to 50%, and wide surgical excision is the most important factor in decreasing the recurrence rate. Chemotherapy and radiation therapy have been effective only in

controlling inoperable lesions and have not been shown to be curative.

9.5.12 Desmoplastic Fibromas

Desmoplastic fibromas are rare bone tumors that many consider the bony counterpart to the desmoid tumor mentioned previously. They are histologically identical. Desmoplastic fibromas do not metastasize but are locally aggressive, requiring wide local en bloc resection.

9.5.13 Langerhans Cell Histiocytosis

Langerhans cell histiocytosis (LCH) is a rare disease involving proliferation of antigen-presenting cells, with clinical presentations ranging from an isolated bone lesion to multisystem disease. More than 50% of patients are younger than 10 years, and most often first present with symptoms of the maxillofacial region. Lesions tend to involve the posterior mandible and clinically present as swelling of the jaw, ulcers, loose teeth, and pain. Radiologic examination typically reveals punched-out radiolucent lesions, with teeth floating in space (► Fig. 9.18). Jaw lesions are present in all three types. Langerhans cell histiocytosis is clinically divided into three groups, unifocal, multifocal unisystem, and multifocal multisystem. Unifocal LCH typically presents with solitary or multiple bone lesions and is treated with curettage, radiation, and interlesional corticosteroids. Overall, it has a good prognosis. Multifocal unisystem LCH is characterized by fever, bone lesions, and diffuse eruptions. When diabetes insipidus is present with exophthalmos and lytic bone lesions, this is called Hand-Schüller-Christian triad. Peak onset is at 2 to 10 years of age, and treatment involves immunosuppressive agents and corticosteroids, with an overall good prognosis. The last type is known as Letterer-Siwe disease

and is found in children younger than 2 years, and the prognosis is poor, with a 5-year survival of 50%.

9.5.14 Burkitt's Lymphoma

Burkitt's lymphoma (BL) is a high-grade B-cell non-Hodgkin's lymphoma and exists in multiple subtypes. The African or endemic variant usually involves the maxilla and other facial bones, whereas head and neck manifestations of nonendemic BL are rare. It primarily affects African children aged 4 to 8 years and is twice as common in boys. Patients often experience swelling, loss of teeth, and mild pain. Treatment involves intensive intravenous chemotherapy and selective surgical debulking. Burkitt's lymphoma is aggressive, with a doubling time of 24 hours, and if left untreated, death occurs in 6 months.

9.5.15 Neurofibroma

Neurogenic tumors are extremely rare within osseous structures in the head and neck region. Neurofibromas are the most common jaw tumors of neurogenic origin. They often present as multilocular radiolucencies and bony expansion. Treatment consists of close observation and surgical excision of lesions that produce pain, paresthesia, or rapid growth. Goal is to excise soft tissue mass and recontour the underlying bone. The tumors are extremely vascular and have a high recurrence rate.

9.5.16 Melanotic Neuroectodermal Tumor of Infancy

The melanotic neuroectodermal tumor of infancy (MNTI) is an uncommon osteolytic-pigmented neoplasm that presents within the first year of life. It primarily affects the jaws, with 95%

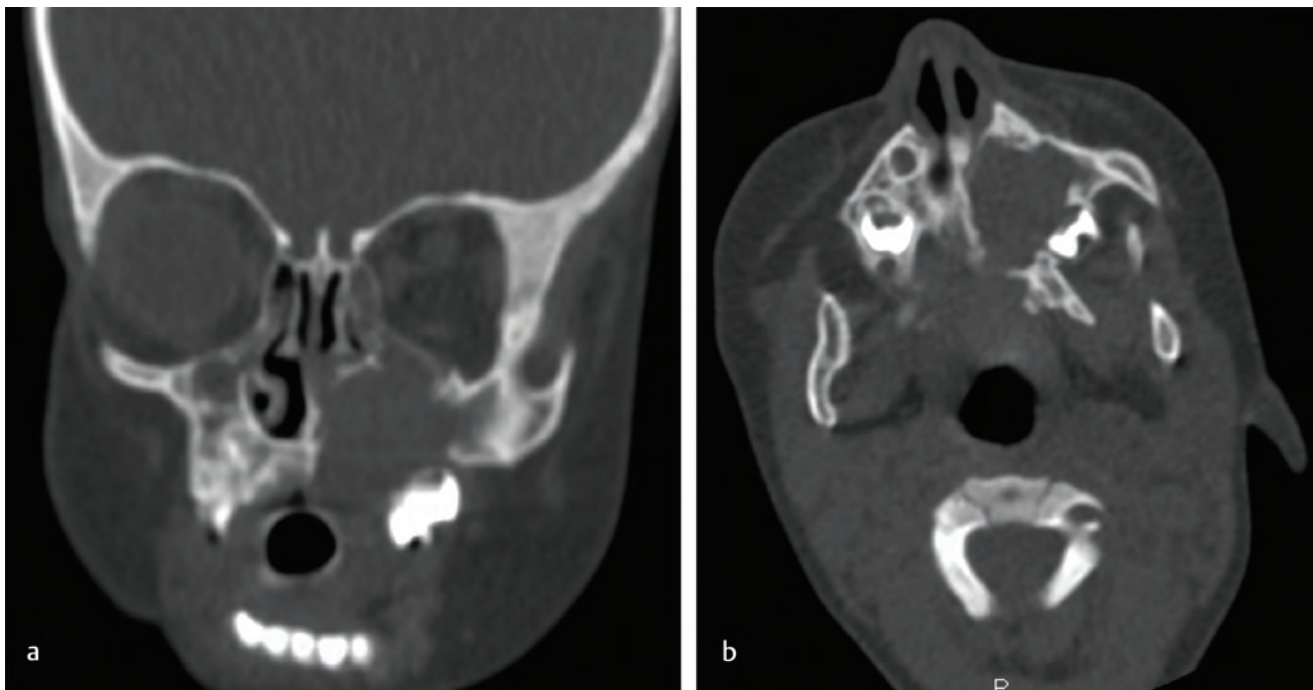
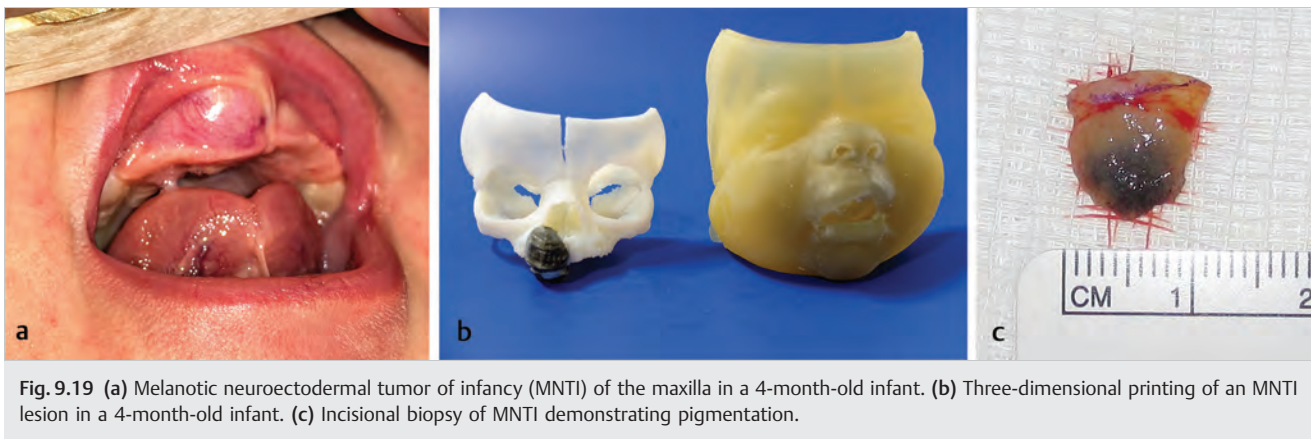


Fig. 9.18 Langerhans cell histiocytosis.



of lesions involving the anterior maxilla (► Fig. 9.19a, b). Similar to other tumors of neural crest origin, such as neuroblastoma, and pheochromocytoma, infants often demonstrate a high urinary excretion of vanillylmandelic acid (VMA). Clinically, the lesions rapidly grow and can often impair sucking and feeding. Often times, there is a deciduous tooth displaced within the lesion. The lesion has a dark melanotic pigmentation (► Fig. 9.19c). Treatment involves surgical excision with 2- to 5-mm margins and is usually curative. Existing teeth and developing buds must be included in the specimen. Recurrence rates have been reported up to 15%. There are some case reports of neoadjuvant therapy reducing the lesion by 50% and should be considered in lesions that will involve the orbit or are inoperable.

9.5.17 Vascular Lesions

Vascular lesions are almost never of primary osseous origin. Intraosseous mandibular arteriovenous malformations are less than 5% of all vascular malformations (► Fig. 9.20a–c). They are more frequent in women and are more common in the head and neck area than extremities. Malformations are usually noticed at birth and grow proportionately but may become clinically relevant when the adult teeth erupt. Teeth may be mobile and pulsatile. Life-threatening hemorrhage may occur with extractions. Management primarily involves interventional radiology for sclerotherapy and embolization (Fig. 9.20e). Primary osseous surgical intervention is rarely indicated.

Malignant tumors involving osseous structures include osteosarcoma and Ewing's sarcoma.

Osteosarcoma is the most common malignant tumor in children, representing more than 50% of all malignant pediatric bone lesions. Less than 5% of pediatric osteosarcomas present in the head and neck area, most often within the jaws (► Fig. 9.21a). It is felt that there is a slight male predilection, and peak incidence is during late adolescence.

The most significant risk factor for head and neck osteosarcoma in children is hereditary retinoblastoma. Other risk factors include previous radiation therapy and certain syndromes that predispose individuals to osteosarcomas such as Li–Fraumeni and Rothmund–Thomson syndromes.

Definitive diagnosis and staging for osteosarcoma require a tissue biopsy, head and neck computed tomography (CT), chest CT, local magnetic resonance imaging (MRI), and bone scan

(► Fig. 9.22b, c). The histologic and molecular profile determines if the lesion is low versus high grade. Management involves local control with surgery and radiation for those patients with positive surgical margins, even though it is felt that osteosarcoma is not radiation-sensitive. For high-grade lesions, adjunctive chemotherapy is often utilized, initially preoperatively as a neoadjuvant, followed by a month for recovery and biopsy to determine the percentage of tumor kill. This is followed by surgical resection and 6 weeks of postoperative chemotherapy. The most significant prognostic factor influencing the overall survival is complete resection with negative margins. Patients with negative margins have a 75% 5-year survival rate compared with 35% 5-year survival rate of those who have positive margins. Unfortunately, bony margins of 3 cm and soft tissue margins of 2 cm are nearly impossible in the head and neck area due to the complex anatomy. The most common site for metastasis is the lungs. However, the regional lymph nodes are rarely involved in metastasis of the tumor, and as a result, neck dissection is not recommended.

9.5.18 Ewing's Sarcoma

Ewing's sarcoma is a destructive and aggressive malignancy of the bone that is rare in the head and neck area. It is mostly seen in patients younger than 20 years and is extremely rare in people of African or Asian descent. Most patients have a painless mass, and up to 20% of patients have metastatic disease at the time of diagnosis. CT imaging shows ill-defined, irregular resorption of bone with focal areas of bone.

Definitive diagnosis and staging for Ewing's sarcoma require a tissue biopsy, head and neck CT, chest CT, local MRI, and bone scan. In addition, Ewing's sarcoma is positive on fluorodeoxyglucose positron emission tomography (FDG-PET), which should also be performed. Finally, bilateral bone aspirates and biopsies are routinely performed for staging in pediatric patients. In addition to traditional formalin fixation, a fresh sample should be secured for cytogenetic testing.

The standard approach to treatment is a multimodality approach with chemotherapy administration and local control either via surgery or radiation therapy. Surgical control involves 3 cm margins and 1 to 2 cm of soft tissue margins. The surgeon should place a plate for reconstruction and delay definitive reconstruction until completion of radiotherapy and chemotherapy.

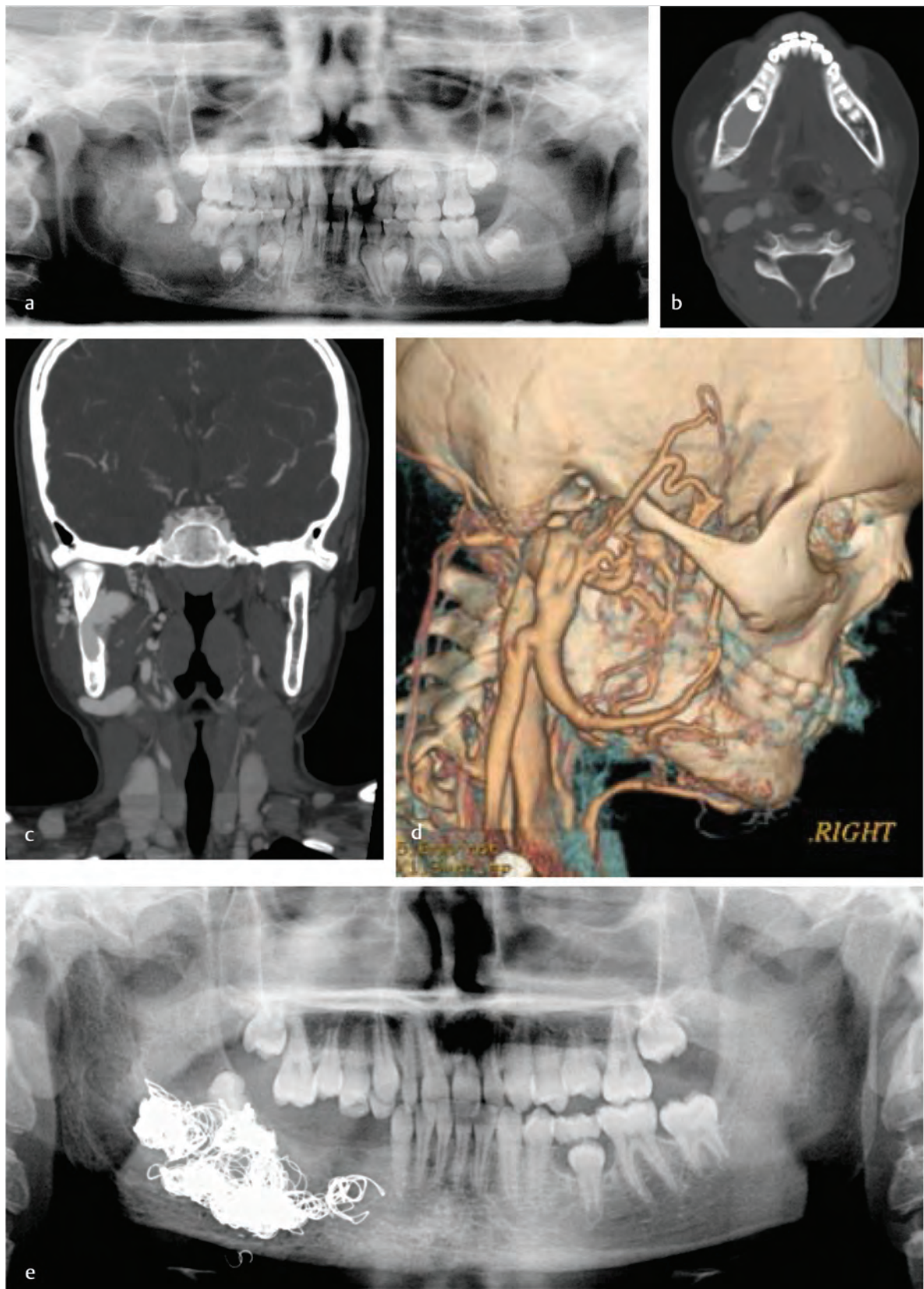


Fig. 9.20 (a–c) Arteriovenous malformation (AVM) of the right mandible. (d) Three-dimensional rendering of vasculature feeding AVM of the right mandible. (e) Postembolization of AVM of the right mandible.



Fig. 9.21 (a) Intraoral view of an osteosarcoma of the right maxilla. (b,c) Osteosarcoma of the right maxilla.

9.5.19 Other Lesions of the Bones

Idiopathic Osteosclerosis

Idiopathic osteosclerosis is a common radiologic finding of asymptomatic, radiodense lesion most commonly associated with vital teeth, found in late first to early second decades of life. If the tooth is nonvital, then the lesion is referred to as condensing osteitis and is thought to be a response to an inflammatory stimulus. The lesions tend to be well-defined, rounded, radiodense masses. They can be managed with radiographic imaging, as they tend to not progress or change into adulthood.

Infantile Osteopetrosis

Infantile osteopetrosis is a rare hereditary skeletal disorder characterized by increased bone density. Bone is unable to be absorbed because of defective osteoclast function, resulting in thick bone and sclerosis of the cancellous bone. When discovered at birth or early in infancy, children often have a diffusely sclerotic skeleton, and often times, there can be evidence of cranial nerve compression. In addition, a common finding is deformity of the face, with frontal bossing, and hypertelorism. Common complications are blindness, facial nerve palsy, and

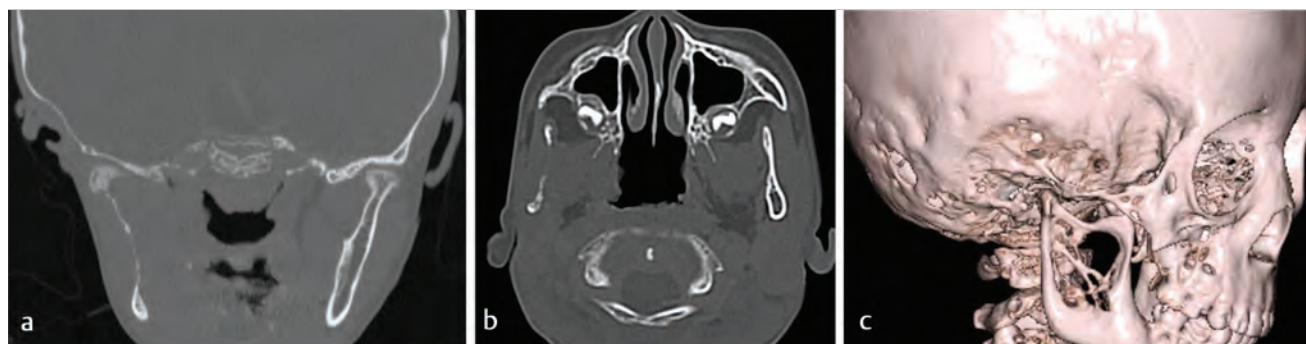


Fig. 9.22 (a) Gorham–Stout disease with subsequent thinning of the right skull base, condyle, and ramus. (b) Gorham–Stout disease involving the zygoma and ramus in the same patient. (c) Three-dimensional rendering of a child with Gorham–Stout disease involving the right skull base, zygoma, TMJ, and ramus.

osteomyelitis secondary to tooth extraction because of tooth disease or delayed tooth eruption. Treatment and management are dependent on the clinical challenge being faced, but in general, care should be taken to minimize potential osteomyelitis and/or fractures of the maxillofacial region.

Gorham–Stout Disease

Gorham–Stout disease (GSD), also known as vanishing bone disease and massive osteolysis, is a rare, poorly understood skeletal condition characterized by spontaneous and progressive disruption of one or more bones. The affected area does not regenerate or repair and fills instead with dense fibrous tissue (► Fig. 9.22a). The disease starts in one bone and may spread to involve adjacent bony and soft tissue structures. Lesions have been reported in the mandible and skull (► Fig. 9.22a–c). Most affected patients are children and young adults. There is often a

history of trauma, and the lesions can occur anywhere in the body. The rate of destruction and extent are quite varied, and there are no predictable factors. Typically, surgical intervention is futile as the process is best managed with radiation therapy and antiosteoclastic medications.

Suggested Readings

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10 Distraction Osteogenesis

Elizabeth Zellner and Derek M. Steinbacher

Summary

Distraction osteogenesis, the gradual repositioning of osteotomized bone in order to generate new bony matrix, is a powerful tool for transforming the craniofacial skeleton. Compared with conventional surgery, distraction may result in shorter operating times, lesser blood loss, and lesser morbidity, along with simultaneous expansion of the associated skin and soft tissue. This gradual approach may overall engender fewer complications and relapse. However, distraction does require longer overall treatment times and significant familial participation. At present, clinical indications for distraction are expanding, and it represents a major tool in the craniofacial surgery arsenal. A thorough understanding of the current science and the applications of distraction osteogenesis is necessary for any pediatric plastic surgery. Three-dimensional modeling and simulation can provide valuable technological adjuncts in planning and executing distraction treatment plans.

Keywords: distraction osteogenesis, mandibular distraction, midface distraction, monobloc, craniostylosis

10.1 Introduction

Distraction osteogenesis is a powerful tool in craniomaxillofacial surgery, and its indications continue to broaden. Gradually repositioning the bones of the craniofacial skeleton entails a smaller initial operation and may mitigate the need for permanent rigid fixation. During osseous expansion, not only new bone is generated but also the soft tissue is influenced and adjacent spaces can be enlarged. Smaller devices with improved mechanics and the advent of three-dimensional (3D) analysis and planning have enabled improved patient experiences and outcomes.

10.2 History of Distraction Osteogenesis

Distraction osteogenesis was first described by Codvilla in 1905, where he described using axial distraction forces to lengthen a femur. This was not adopted into mainstream medicine until Ilizarov, a Russian physician, popularized the technique in the lower extremity starting in 1949. The technique was vetted in a series of physiologic experiments, where bone generation was confirmed following gradual traction to the callus. The functional load on bones and their blood supply also showed impact on morphology and articulation points. In parallel, steady traction also encouraged the accommodating expansion of the adjacent soft tissue, including periosteum, blood vessels, ligaments, cartilage, muscles, and overlying skin, known as distraction histogenesis.

In the 1970s, Snyder described lengthening of the canine mandible by using a similar method of bony distraction. This technique was then applied to humans in the early 1990s, to

treat patients with Nager's syndrome and hemifacial microsomia. Since that time, additional craniofacial applications have taken advantage of this powerful tool for gradually lengthening the skull and facial bones, along with the overlying soft tissue.

Throughout the intervening decades, distraction has been used, with significant success in the mandible, and also for alveolar distraction, Le Fort I maxillary advancement, Le Fort II and III midface advancement, monobloc/fronto-orbital advancement (FOA), and cranial vault expansion. As each new technique has evolved, a more thorough understanding of the uses and limitations of distraction has also become more apparent.

10.3 Principles of Distraction

The principle of distraction osteogenesis is straightforward. The procedure begins in the operating room with a carefully planned osteotomy across the region where bony expansion is desired. This osteotomy may be full or partial thickness, depending on the adjacent structures present, and the orientation of these cuts is critical to achieving the desired result. Once the osteotomy is made, a distraction device (internal or external) is applied with grounding footplates or pins on either side of the osteotomy. The vector of this device placement determines the 3D direction of the distraction and bony regenerate formation. Depending on the age of patient, time may be given for a latency period, where the device is held in its original position, with no forces applied across the devices. Once distraction begins, the device is activated and it stretches the span across the osteotomy site. This is done in a slow, controlled fashion, which enables the regenerated bone matrix to form across the osteotomy as it is stretched. Once the desired end position of distraction is reached, the devices are left in place for consolidation. This time period permits the bony regenerate to ossify and strengthen into true new bone. After this, the devices can be removed and the newly generated bone will maintain the distracted length (► Fig. 10.1).

The advantages of distraction osteogenesis include the gradual distraction histogenesis, first described by Ilizarov. Since the two sides of the osteotomy are not extensively manipulated in the operating room, there is less immediate trauma to the tissue, which means less blood loss, less swelling, and shorter operative and hospital admission times. Unlike osteotomies with repositioning and bony fixation immediately in a new location, there is no need for bone grafting, as the bone incrementally forms itself. Distraction may also mitigate future growth restrictions by avoiding need for wide subperiosteal exposure and fixation. With external devices, there is also the opportunity for midcourse vector correction if the tissues do not respond exactly as predicted to the distraction plan. In addition, distraction may be appropriate for smaller children who may not tolerate single-stage bony movement.

Disadvantages of distraction pertain to the hardware requirement and total treatment time. Patients and families are educated that this is not a one-and-done surgery but instead

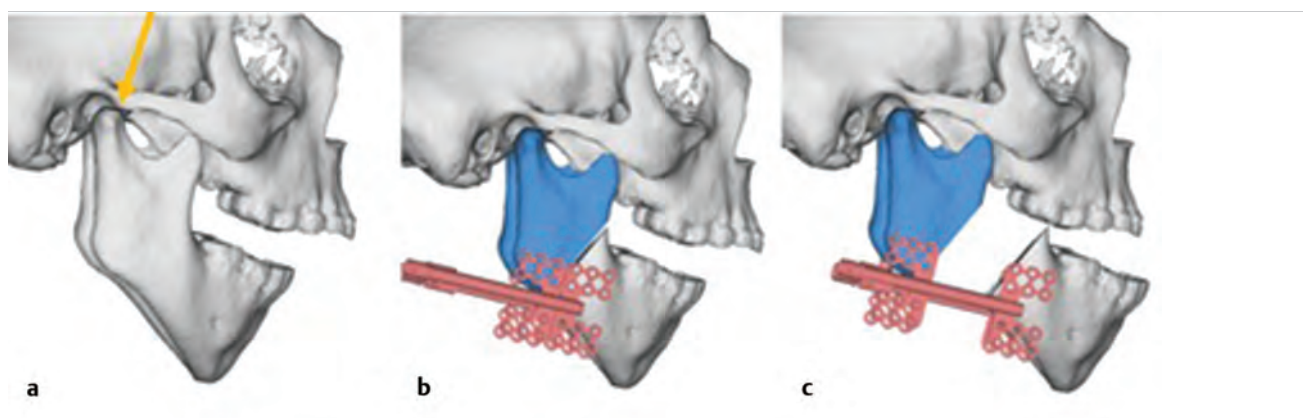


Fig. 10.1 Three-dimensional plan showing (a) hypoplastic mandible before distraction osteogenesis, (b) mandibular osteotomy perpendicular to plane of planned distraction, with internal device oriented along planned distraction vector. Please note the inferior fixation plates would be removed during actual surgery. (c) Simulated position of mandible following planned distraction. Demineralized bony regenerate will form along the gap during the activation period and will mature into calcified bone; however, the device stays in place during the consolidation period. Once this is complete, the device will be removed, with newly formed bone spanning the distraction length.

occurs over a period of weeks to months and requires activation and cooperation by the caretakers. The distraction devices, which may remain on for several months, may be distressing to parents if not adequately prepared. Often, much of the distraction is carried out at home, which requires less hospitalization time, but it does require participation of parents to turn the distraction devices and accurately record the protocol. Distraction procedures require a strong treatment relationship and communication between the craniofacial teams and the patients/families under their care, in order to reach a successful outcome.

10.3.1 Scientific Principles of Distraction Osteogenesis

During distraction osteogenesis, a number of critical events are carefully orchestrated at the molecular level. During the distraction phase, bone forms in response to the forces of tension and stress on the regenerate, which heals by membranous ossification. The collagen fibers are oriented parallel to the direction of distraction, and osteoid is deposited in the center of the regenerate. Ossification then advances from either end of the central fibrous zone, which forms an immature bone bridge across the distraction defect. This immature bone then remodels, beginning in the consolidation phase and continuing for 1 to 2 years. Although the regenerate is never as strong as the native bone, it eventually reaches approximately 75% of native bone strength.

Limitations of distraction osteogenesis may occur in patients who have poor local and systemic substrates (► Table 10.1). In addition, devices may become dislodged by trauma or infection, which can result in malunion or nonunion. Longer periods of consolidation may also be cumbersome to some patients. As a result, much recent research has been aimed at increasing bone deposition to possibly shorten the overall treatment time. Rat studies have used adenovirus-driven transcription of the gene for *BMP-2* to enhance bony deposition. It is also known that ischemia can be a limiting factor in distraction. Studies have

Table 10.1 Local, systemic, and distraction factors that may affect the outcomes in distraction osteogenesis

Local factors	Systemic factors	Distraction factors
Osteoprogenitor supply Blood supply Infection Soft tissue scarring Bone stock Radiation history	Age Metabolic disorders Vitamin D deficiency Connective tissue disease Steroid therapy Calcium deficiency	Latency period Rate of distraction Rigidity of fixation Consolidation period Regenerate length

shown that endothelial progenitor stem cells are drawn to this ischemic site during activation and they remain through the consolidation period. These stem cells may represent another possible source to overcome ischemic limitations of distraction. Cyclic mechanical strength has been related to expression of *IGF-1*, *TGF- β* , and *bFGF* along with interleukins (IL)-1 and IL-6, possibly the effectors of further bone growth in distraction osteogenesis. Numerous interventional cytokines have been utilized to optimize bony deposition and possibly decrease overall treatment time. A list of these substances can be found in Box 10.1.

Box 10.1 An Overview of Some Molecular Cytokines' Interventions and Their Effect on Distraction Osteogenesis

- Bone morphogenic protein
- Endothelial progenitor cells
- Alkaline phosphatase
- Transforming growth factor- β 1
- Vascular endothelial growth factor
- Basic fibroblast growth factor (bFGF)
- Interleukin 1
- Interleukin 6

Table 10.2 Phases of distraction osteogenesis and considerations during each phase

Phase	Osteotomy	Device placement	Latency	Activation	Consolidation	Device removal	Growth modification
Time	0	0	0–7 d	Days–weeks	Months (at least 6 weeks or 2 × activation)	End of treatment	After device removal
Considerations	Full thickness Adjacent structures Orientation	Orientation Number	Age of patient Anatomic area Membranous vs long bone?	Age of patient Anatomic area Length desired Device length	Age of patient Device complications Infection risk	Reossified bone Timing Retaining hardware	Orthodontic appliances, elastics

10.3.2 Distraction Protocols

Distraction osteogenesis consists of three general phases: latency, distraction, and consolidation (► Table 10.2). The first stage, latency, occurs after bony osteotomy but before activating the distraction device. During this time, the surgeon allows preliminary callus to form but does not want the regenerate (the intermediate stage of bony deposition) to begin to reossify through calcification. Depending on the bone being distracted, those skeletal components with a richer blood supply or membranous origin (such as the mandible and the skull plates) require shorter latency periods. In addition, younger patients, such as neonates and young infants, may not require any significant latency period before distraction. For mandibular distraction in these patients, the surgeon can either start distracting immediately or wait only 24 hours to begin distraction. In contrast, older patients, who are skeletally more mature, may require a slightly longer latency period, usually 3 to 5 days when distracting the mandible.

Distraction is the phase in which the physical lengthening of the bone is instigated through active manipulation of the distraction device. Using the distraction device, the callus that is formed on either side of the osteotomy site is stretched gradually. During this time, there is exuberant angiogenesis. This vascularized osteoid stimulates new bone formation across the osteotomy defect, which forms parallel to the distraction vector.

Much has been written on the ideal frequency and rate of distraction, and there is significant variation among different authors and practitioners. There must be a balance between distracting too fast, which results in a fibrous nonunion, and distracting too slow, which can cause premature bony union that prevents further lengthening. Perhaps, the most frequently cited rate of distraction is 1 mm/day, generally separated into two daily increments of 0.5 mm. However, distraction rates of 2 mm/day and even 3 mm/day have also been reported. Younger patients, such as neonates, can generally tolerate more rapid distraction, as their bony healing patterns are more robust. The indications of airway creation to avoid a tracheostomy or of cranial expansion to allow brain growth in these patients also generally provoke a more urgent, more rapid course of distraction.

The final phase of distraction osteogenesis is consolidation. During this period, the device is used to maintain the physical lengthening achieved by the distraction period. While the bone is being stabilized in this manner, the bony regenerate can reossify through calcification into a mature bone. The consolidation phase is generally twice as long as the active distraction phase

Table 10.3 Types of devices used for distraction osteogenesis

Device type	Pros	Cons
Internal	Comfort for patient and family Less obtrusive	Infectious risk Unable to adjust vector Second removal operation
External	Vector adjustable Single or stage	Cumbersome to patient and family
Linear	Straightforward Device fixation	Limited dimensions
Curvilinear	More anatomic	Complex planning Challenging engineering
Multivector	Precise control Multiple dimensions	Complex planning Require multiple devices Device fixation

to allow sufficient bone maturation, so that the device is no longer needed for structural stabilization. Consolidation periods of 4 to 12 weeks have been cited, but a period of at least 8 weeks is preferred and has been shown to generally prove sufficient in most cases. Again, younger patients generally require shorter consolidation phases because of their faster healing. Load-bearing bones, such as the mandible, may require longer consolidation periods. When in doubt about an appropriate consolidation period, ultrasound or radiographs can be used to check on the progress of bony remineralization.

10.4 Devices

To achieve these goals, a number of different devices have been used throughout the years (► Table 10.3). Initially, external distractor devices utilized transcutaneous pins to fixate either side of the osteotomy site with external distraction arms to exact the lengthening vector (► Fig. 10.2). These devices, with their active parts exposed and available for manipulation, allow for alteration of the distraction vector during the active distraction period. The obvious downsides of these devices include significant bulk and unsightly pin tracks in the skin. Since then, internal devices have been developed. These are placed directly on the bone under the skin and soft tissue; however, periosteum is ideally left intact on at least one side of the osteotomy site to guide the formation of the bony callus. Internal devices also include a distraction arm, which comes through the skin, usually at some length away from the device to discourage

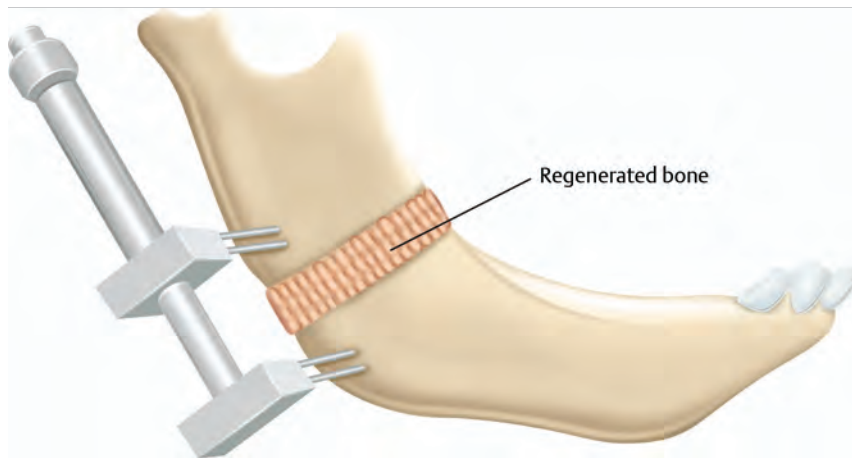


Fig. 10.2 Initial studies utilized external distractors that were inserted percutaneously into bone.



Fig. 10.3 Internal distractors were subsequently developed with vector arms through which to transcutaneously submit distraction vectors.

infection (► Fig. 10.3). This arm is manipulated to activate distraction of the device. Internal devices can be concealed in a more aesthetic fashion and can be more tolerable to patients and their families, with fewer external components to dislodge. This may permit for longer consolidation periods. However, these can be difficult to securely fixate in very young bone, they must be removed at a second operation, and the vector of internal devices cannot be manipulated during the distraction period.

Additional relevant factors include device placement to determine the vector of distraction. In addition to the orientation of placement, devices can be uni- or multidirectional, which will influence the course of distraction based on clinical objectives. Curvilinear devices are also available and implemented in certain contexts (► Fig. 10.4). Many variations of footplates, device arms, and morphologies exist, customized for a wide range of applications.

Resorbable distraction devices have been designed and implemented in patients for many types of distraction osteogenesis. These devices hold the promise of single-stage (or much smaller second stage) distraction protocols that hold the bones in consolidation for a longer period of time but never need to be removed. In addition, programmable, automated devices have been investigated and successfully used in human mandibular distraction osteogenesis in an adult patient.

Although these devices have not yet gained widespread use, expandable metal alloys, springs, and pneumatic controls may make distraction protocols simpler for patients and their families in the future.

Three-dimensional computed tomographies (CTs) and software planning tools offer improvement to distraction planning and application. An initial CT scan allows a quantitative measurement of the morphology and the existing bone–soft tissue relationships. Computer-assisted design and manufacturing (CAD/CAM) permits virtual surgical osteotomies, device placement, and proposed movements to optimize the clinical result. Three-dimensional prints based on the simulation can then translate the digital plan intraoperatively with cutting guides and templates (► Fig. 10.5). Some have even advocated image-guided and robotically performed osteotomies to further enhance precision in delivering a 3D computer-aided surgical plan.

The distraction plan must incorporate a number of factors. The dysmorphology and deformity must be accurately analyzed, and the goals of correction (size, length, direction, and magnitude) must be accurately defined. Future growth and overcorrection must also be considered. The device type, direction, and placement must be carefully selected and individualized to the patient. The distraction protocol is then discussed with the patient or family at length before

beginning distraction, as success is incumbent upon their participation. Consolidation and other retention measures are implemented to maintain the distracted outcome goals with time.

10.5 Specific Applications for Distraction Osteogenesis

10.5.1 Mandible

Mandibular distraction is the most common application, typically for expanding the pharyngeal airway and alleviating glossoptosis. This is indicated for airway compromise that may otherwise require a tracheostomy. Both syndromic (i.e., Nager's, Treacher Collins, or Stickler's) and nonsyndromic (Pierre Robin's sequence) patients may benefit from distracting the mandible. Mandibular distraction is considered if other sites of airway obstruction are minimal or exonerated, for patients requiring persistent intubation/tracheostomy (or the desire to decannulate these patients), for an airway that cannot be managed by conservative measures such as prone positioning, or if sleep studies indicate moderate to severe obstructive sleep apnea (► Table 10.4; ► Fig. 10.6).

Internal and external distractors can be used with a variety of vectors. New-generation internal devices are small enough to accommodate a neonatal mandible and are preferred. Distraction vector depends on surgeon preference, without broad consensus. Multivector distraction is touted to mimic normal morphology and growth, especially in older children. However, a single-vector orientation is often used in small, infant mandible. Bilateral bicortical osteotomies are usually performed posterior to the developing tooth roots, in an oblique or vertical fashion, taking care to preserve the facial nerve and inferior alveolar nerve branches. The devices should be placed parallel and collinear, taking into account the inferior border, adjacent anatomic structures, and planned trajectory. For micrognathia or retrognathia causing airway obstruction, the primary



Fig. 10.4 An X-ray showing a curvilinear in place to recreate a more complex anatomical vector in an older patient.

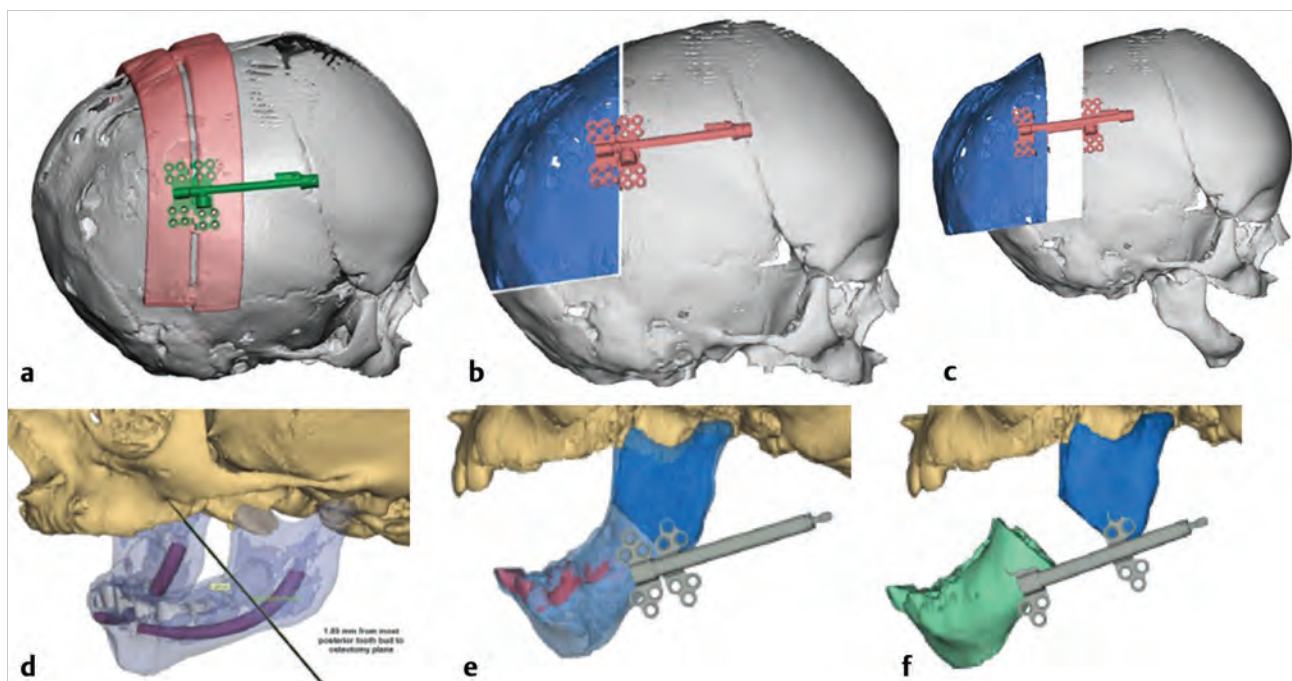


Fig. 10.5 Three-dimensional plans from CT scans utilized in posterior vault distraction. (a–c) Computed tomographic images can be used to assess bony, soft tissue, and intracranial anatomy and print custom cutting guides to precisely and efficiently affect the surgical plan in the operating room. In mandibular distraction, the course of the inferior alveolar nerve and tooth buds can be analyzed with imaging to assist surgical planning (d–f).

Table 10.4 An overview of distraction osteogenesis in the craniofacial skeleton

Anatomic Area	Indications	Benefits	Downsides
Mandible	Retrognathia (airway) Vertical length (asymmetry) Proximal ramus/TMJ	Bone generation Alleviate glossoptosis Vertical mandibular length	2 procedures
Maxilla	Le Fort I (cleft maxilla) Large magnitude maxillary movements	Achieve large sagittal advancements Less relapse Speech	2 procedures Occlusion
Alveolar	Vertical length Medial/lateral (cleft)	Avoid or less bone graft for dental implants, or close alveolar fistula	2 procedures
Midface/monobloc/ bipartition	Syndromic midface and frontal hypoplasia	Achieve large advancements Less relapse Less dead space or infection	2 procedures Cumbersome External device Occlusion
Cranial	Syndromic synostosis Chiari	Gradual vascularized expansion Less relapse	2 procedures
Transport	Bone defects	Close critical-sized defect with autologous tissue	2 procedures

Abbreviation: TMJ, temporomandibular joint.

distraction goal is to enlarge the retroglossal airway. Ideally, this is done to preserve normal mandibular morphology and to avoid occlusal malrelationships, but the airway is the prime outcome variable. Pulling the tongue anteriorly, and also inferiorly, has the greatest influence on the posterior airway space. Both sagittal and oblique vectors can accomplish airway enlargement with excellent clinical outcomes. Mandibular distraction is effective to avoid tracheostomy and allow decannulation in both infants and older children with airway obstruction secondary to retrognathia.

Combined vertical ramus and horizontal mandibular hypoplasia in hemifacial microsomia and other causes of asymmetry can also be addressed using distraction (► Fig. 10.7). External devices were used in early generations to vertically and coronally improve the mandibular morphology. The Pruzansky/Kaban's classification of mandibular hypoplasia differentiates the degree of mandibular hypoplasia based on the degree of mandibular ramus and temporomandibular joint involvement. Type I and II mandibles are generally amenable to ramus distraction in a largely vertical vector to correct the mandibular asymmetry. In type III mandibular hypoplasia, the proximal ramus–condylar unit is entirely absent, with no articulating temporomandibular joint. An intact proximal stop in the form of a functional temporomandibular joint is a prerequisite for mandibular distraction. In these severe cases, replacement of the ramus will be first necessary, often with either free rib graft or free fibula flap, which may then require further lengthening with distraction osteogenesis.

Controversy about the progression of asymmetry in hemifacial microsomia (HFM) exists, and one reason to intervene with distraction during the mixed dentition is to avoid worsening of the deformity. Other evidence suggests that the asymmetry remains proportional with time and growth and that early distraction may be subject to relapse. However, orthodontic bone anchorage, bite wafers, and growth modulation may help

maintain postdistraction form. Early intervention may also be indicated for psychosocial issues, and the type IIB or III mandible may benefit from ramus reconstruction with subsequent distraction. For type I and IIA cases, if distraction is deemed appropriate, then age 6 to 8 years is chosen both for the frequency of peer interactions and to take advantage of permanent tooth eruption to help level the frontal occlusal cant. A posterior open bite should not be left to close on its own, but rather, encouraged passive or active eruption, while maintaining the mandibular position, is employed. Distraction is typically not purely vertical in these patients but rather a combination, as the deformity is in three dimensions.

10.5.2 Le Fort I

Distraction has also been used in the maxilla for midface deficiency in cleft and syndromic patients (► Fig. 10.8). It has been estimated that between 25 and 60% of patients born with unilateral cleft lip and palate will require maxillary advancement to correct functional deficits and unsatisfactory facial aesthetics due to maxillary hypoplasia. Maxillary hypoplasia has been associated with speech and feeding difficulties as well as with issues relating to nasopharyngeal airway patency. Traditionally, very large Le Fort I advancements, such as those greater than 10 mm of anteroposterior advancement, and significant advancement in cleft patients have been associated with increased relapse.

After observing success in mandibular distraction, facemasks, interdental splints, and then internal devices were used to advance the maxilla gradually following Le Fort I osteotomy. Although the initial external distractors were sometimes decried for being cumbersome, significant progress has since been made with internal devices, even when maxillary advancements greater than 10 mm are indicated. Distraction can achieve greater advancement than conventional, one-stage



Fig. 10.6 Mandibular distraction done for mandibular hypoplasia in order to facilitate tracheostomy decannulation in an older patient (a,b) and to avoid tracheostomy in an infant with isolated Pierre-Robin sequence (c-e). Pre- and postoperative 3D CTs of a patient undergoing mandibular distraction (f,g).

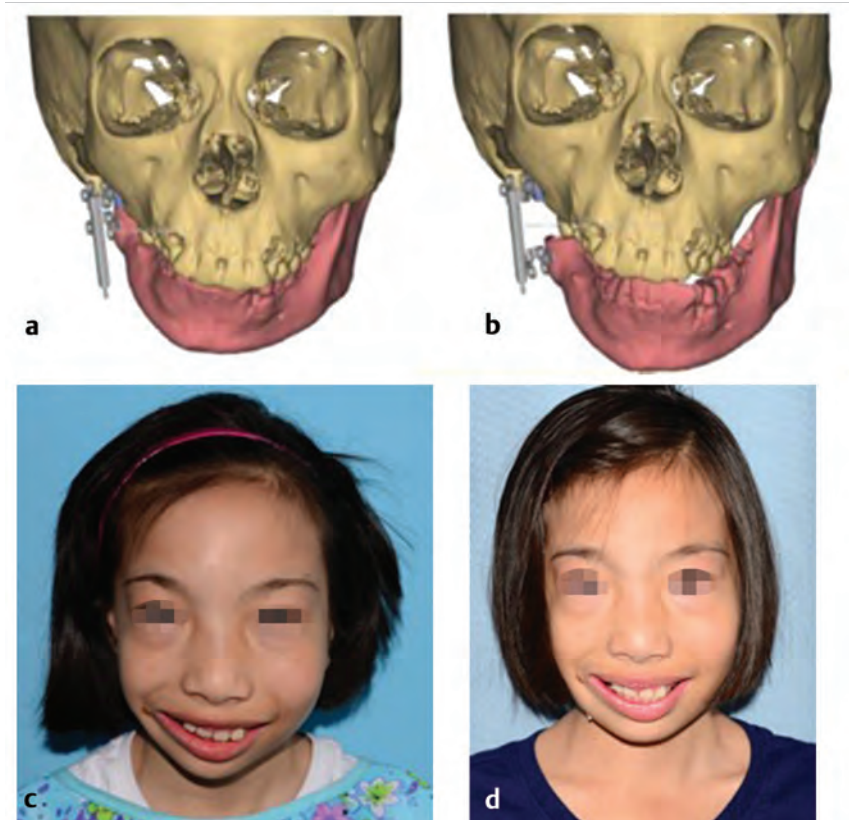


Fig. 10.7 Mandibular distraction osteogenesis CT planning (a,b) and pre- and postoperative photographs (c,d) in a patient with hemifacial microsomia.

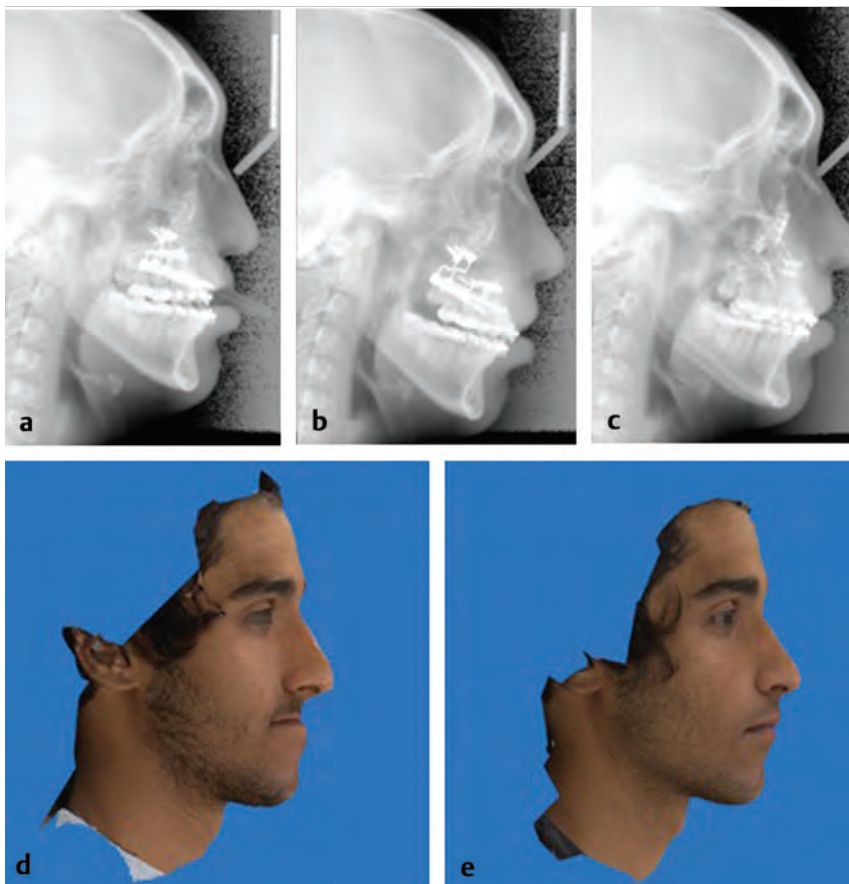


Fig. 10.8 Maxillary distraction in cleft patients has been effective, with less relapse than conventional surgery. Pre- and postoperative cephalograms (a–c) and 3D photographs of a patient with a history of cleft palate who underwent maxillary distraction (d,e).

Le Fort I movements and may minimize the need for mandibular setback, which can compromise posterior airway space. Though not routinely used, maxillary distraction may be considered in young patients with severe maxillary deficiency. Studies have also shown decreased relapse and improved speech outcomes in cleft patients who undergo distraction osteogenesis versus traditional maxillary osteotomy and plating. A hybrid approach may be employed, where the maxilla is distracted to length, and rather than implementing a consolidation phase, the devices can be removed and the maxilla plated. Distraction devices have also been indicated in the orthodontic literature to close large alveolar clefts, with some success.

Midface

Frontofacial and midface distraction osteogenesis has been gaining more support in recent years, usually in patients with syndromic craniosynostosis, such as Crouzon's and Apert's syndromes. Computed tomographic morphometry has elucidated that although these patients have similar midface bony volume compared with normal controls, they have altered sphenoid morphology, with diminished growth inferiorly and anteriorly. Large anterior and inferior movements can be required to address significant midface issues of exorbitism and nasopharyngeal airway and could cause considerable morbidity, with likely relapse in a single-stage operation. In addition, with a single-stage procedure, a complete frontofacial procedure, such as a monobloc, leaves a large dead space that is in communication with both the cranial vault and the nasal passages, presenting a concerning source for infection. In contrast, gradual distraction obviates the formation of this dead space as the soft tissue stretches in parallel with the bony movements. Frontofacial

advancement with distraction osteogenesis has been shown to produce stable, satisfactory results in multiple studies.

Most distraction of the midface follows the conventional Le Fort osteotomies shown by Tessier. Later, the monobloc was described by Ortiz-Monasterio. It moves the midface and frontal bone as one piece and was then distracted with success by Polley et al. Although there are many options for midface distraction, the individual osteotomies and movements must be modified and the facial skeleton individually partitioned to address the specific needs of the patient.

10.5.3 Le Fort II and III

The Le Fort III segment is distracted for syndromic midface deficiency, where the orbits, nose, zygoma, and maxilla are all deficient. The orbits and upper airway are enlarged, improving globe position and protection as well as airway obstruction (► Fig. 10.9). In addition, the malar region is made more prominent, the nasal morphology is improved, and occlusal conditions are optimized. Either internal or external devices can be used in this area, both with success and proponents. Longitudinal follow-up of these patients shows a trend toward greater advancement, with less relapse with Le Fort III distraction versus conventional Le Fort III. This may assist in lowering the number of operations in patients who otherwise would have a significant number of surgeries throughout their lifetime.

Hopper et al have proposed a differential midface repositioning using a Le Fort II distraction with zygomatic repositioning for midface advancement in patients with Apert's syndrome. Through this technique, they hope to address the syndromic central facial concavity, in addition to its malposition. Although

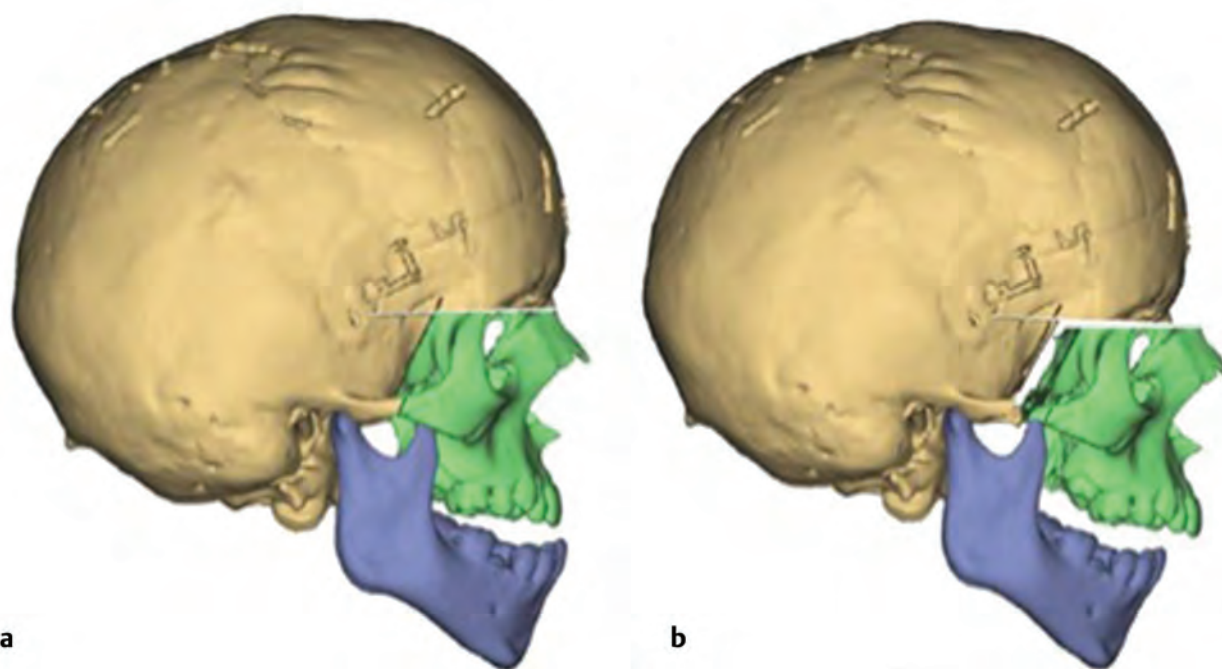


Fig. 10.9 Le Fort III 3D CT advancement plan for a patient with Crouzon's syndrome (a,b).

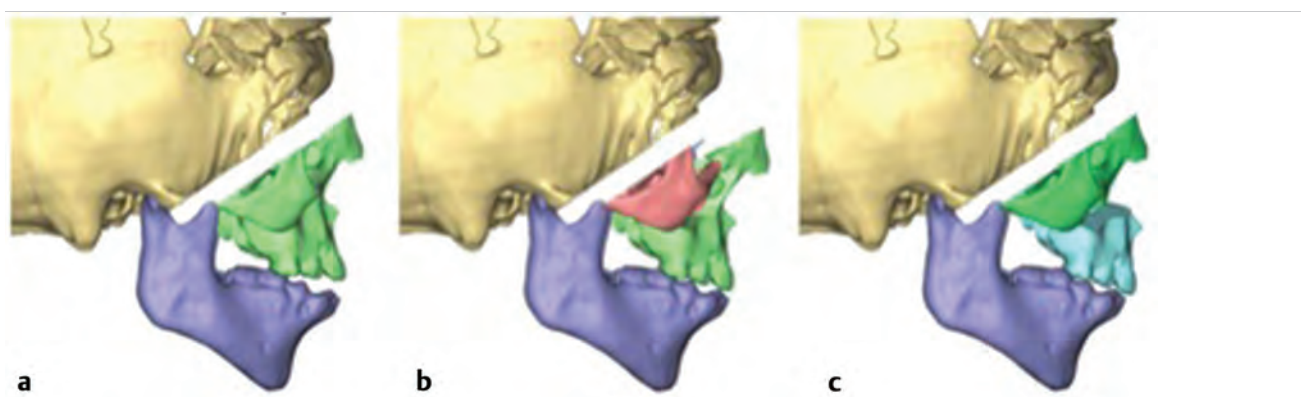


Fig. 10.10 A number of different surgical plans can be utilized to improve skeletal outcomes in patients. (a) Le Fort III osteotomy. (b) Le Fort III osteotomy with zygomatic repositioning. (c) Le Fort I osteotomy with Le Fort osteotomy.

there is less literature on advancement using a Le Fort II osteotomy, this distraction pattern combined with simultaneous zygomatic repositioning showed improved normalization of facial ratios on CT morphometry. Customizing the osteotomy plan to an individual patient's morphology, with guidance from 3D CT morphometry, may represent the future for improving outcomes in these complex patients (► Fig. 10.10).

Monobloc

In cases where both the midface and the fronto-orbital region are deficient, monobloc distraction may be considered (► Fig. 10.11). In this circumstance, the midface and frontal bones are moved in conjunction to address both the forehead and the midface. This approach allows for the most complete resolution of exorbitism but imparts communication of the intracranial and nasal/oral cavities. A single-stage monobloc with significant advancement results in an unfavorable complication profile. Patients with ventriculoperitoneal shunts had the highest complications due to a lack of brain expansion, to fill the extradural space, following surgery. For conventional surgery, a two-staged (FOA and Le Fort III) method exhibited less morbidity compared with monobloc. Kawamoto compared conventional with distracted monobloc, indicating a decrease in complications from 61% to 8%, including meningitis, cerebrospinal fluid (CSF) leak, bone flap loss, and wound infections. The distraction group also had less blood loss, a shorter inpatient hospital stay, and larger advancements.

Therefore, when indicated, a monobloc should be performed using distraction osteogenesis exclusively. Polley et al first described using distraction osteogenesis in conjunction with a monobloc osteotomy, using an external distraction device. Internal devices can also be used for monobloc distraction. Distraction has reintroduced a powerful tool in the monobloc, with fewer adverse effects. Facial bipartition may also be used in conjunction with monobloc advancement to help with midface concavity in syndromic patients. Outcomes analysis of midface monobloc distraction has been satisfactory in numerous studies, with greatly improved safety profiles.

10.6 Neonatal Midface Distraction

Neonatal midface distraction has been used in extreme cases of exorbitism, especially in Pfeiffer's syndrome. In 1983, Muhlbaier et al described conventional monobloc advancement in a newborn but cautioned that this should be used only in cases of severe functional impairment. Polley et al first described monobloc distraction in a newborn with Pfeiffer's syndrome and severe exorbitism. Initially, there was enthusiasm for early intervention in this patient, but this has dimmed after significant relapse was observed, which required further surgery. If possible, supportive therapies such as tarsorrhaphies and even tracheostomies may present better alternatives to support a young infant for many months/years until the child's bone stock and ability to tolerate major surgery may be improved. However, Dunaway's recent review of personal experience in very young children (average, 18 months) treated with monobloc for functional issues showed successful resolution of symptoms. The proper lower cutoff for utilizing monobloc advancement remains a highly contentious issue in craniofacial surgery.

10.7 Posterior Vault

Posterior vault expansion through distraction osteogenesis has changed the treatment sequence paradigm for craniosynostosis syndromes (► Fig. 10.12). Although traditionally, the anterior vault was the first target of surgical intervention, relapse and lack of growth necessitate a high rate of redo FOAs later in childhood in this population. In contrast, first expanding the posterior vault enlarges the intracranial volume largely and allows for delay of the FOA to a later date, in the hope of requiring the FOA only once. In addition, compared with conventional posterior vault remodeling, the modification of distraction enables a larger expansion, with soft tissue accommodation, and less relapse associated with supine head placement. Distraction also imparts the benefit of leaving the inner table attached to dura, maintaining vascularization of the posterior cranial segment and enabling concurrent "brain" expansion with distraction. The decreased dissection also means decreased risk to the torcula and posterior venous system, and the reduced dead space may limit the risk of collection and infection.

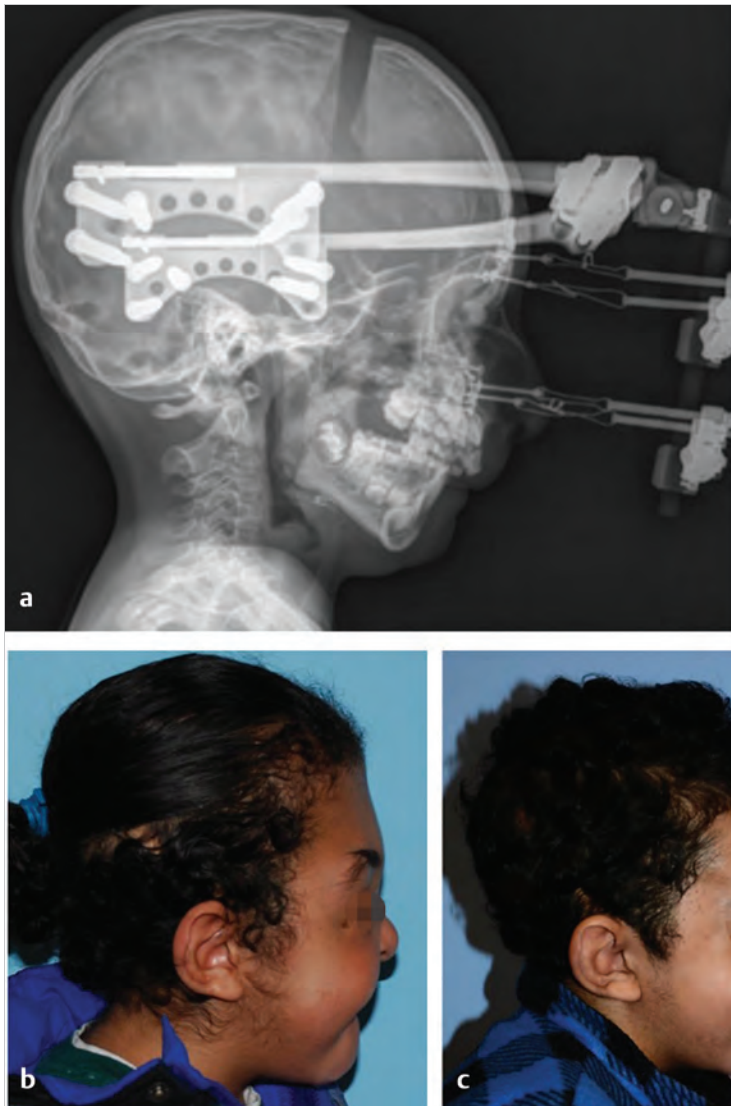


Fig. 10.11 Monobloc advancement of the face, including the frontal bone and the midface in one concerted movement, was first described by Ortiz-Monasterio. Monobloc distraction moves the frontal bone in conjunction with the midface but cannot increase nasal length. (a) Lateral cephalogram of a child undergoing monobloc advancement. (b) Preoperative photograph of a patient with Apert's syndrome. (c) The same patient following monobloc distraction osteogenesis. Orbital protection and airway obstruction were greatly improved following the procedure.

The human skull reaches approximately 77% of its adult volume by 2 years of age and 90% by 5 years of age. In normal children, the majority of this initial growth occurs in the posterior fossa, whereas in syndromic patients with bicoronal craniosynostosis, expansion occurs in the middle cranial fossa, with an underdeveloped posterior fossa. Posterior distraction addresses this discrepancy anatomically, helps normalize morphology, and limits turribrachycephaly. The frontal region may experience added projection as an indirect effect of distraction, based on Newton's third law. In addition, Arnold-Chiari malformations may be simultaneously addressed.

10.8 Anterior Cranial Distraction

Traditional fronto-orbital advancement with fixation remains the gold standard for treating anterior cranial constriction. However, several groups have reported on experimental success with anterior cranial vault distraction. The data are currently limited and without longitudinal follow-up, but theoretic benefits may exist, including maintenance of vascularity. Critics of

anterior cranial distraction cite the effectiveness of conventional techniques, without technical difficulty, and, perhaps, suboptimal correction of turriccephaly with fronto-orbital distraction. There are also concerns about postoperative stability and the risk of reoperation in these patients with more scar tissue present. A more recent direct comparison between conventional FOA and distraction for cranial remodeling failed to show any difference in anthropometric outcomes between groups. However, frontal distraction may gain traction as an option for forehead deformities in the future.

10.9 Bone Graft and Flap Distraction

In addition to the native bones of the craniofacial skeleton, various reports have shown that distraction can be applied to heterotopic bone from both avascular bone grafts and microvascular free bone flaps to increase the utility of this transplanted tissue (► Fig. 10.13). The grafts and flaps are allowed to incorporate and fully heal before distraction is applied.

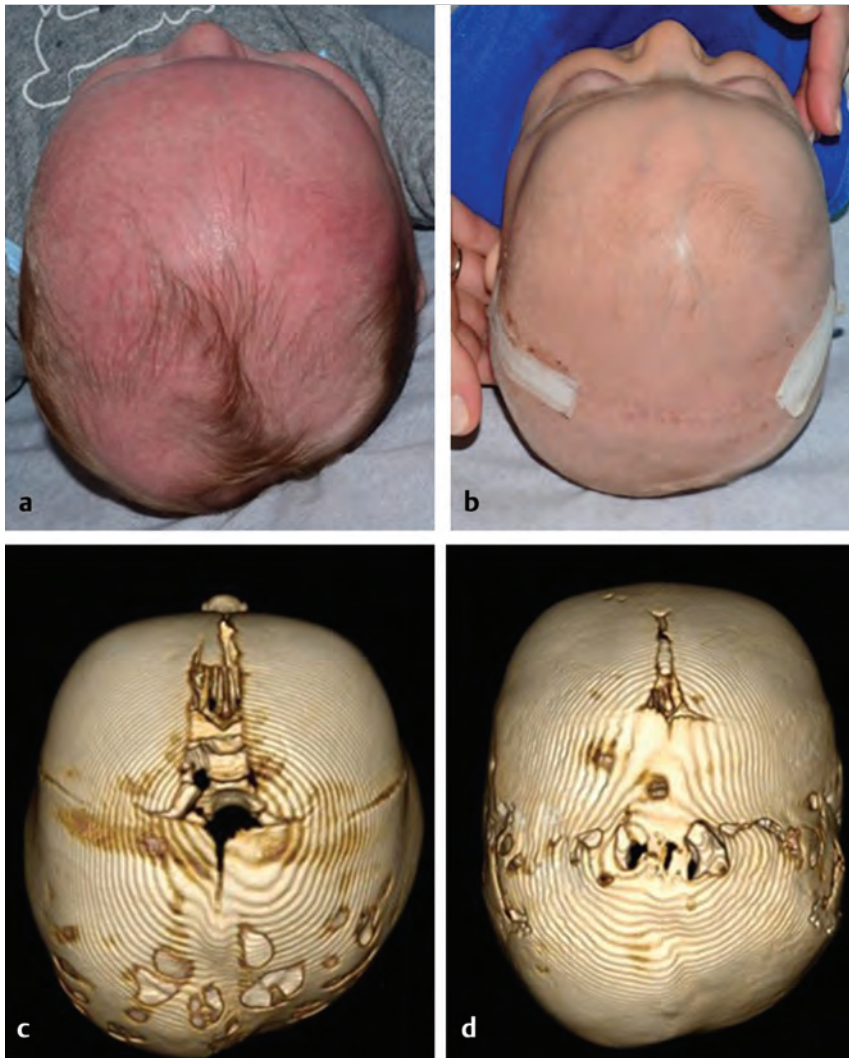


Fig. 10.12 (a) Pre- and (b) postoperative photographs of a patient with Pfeiffer's syndrome, bicoronal craniosynostosis, and resulting brachycephaly, who underwent posterior vault distraction osteogenesis with (c) pre- and (d) postoperative 3D CT scans.

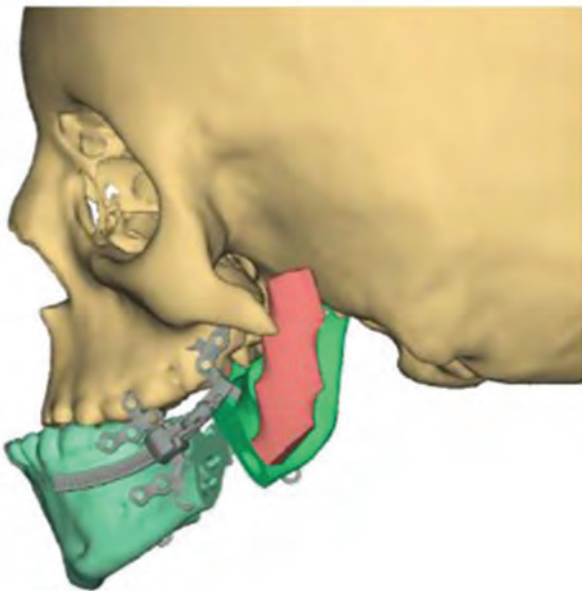


Fig. 10.13 Reconstruction of a Pruzansky III mandible required rib graft and eventual distraction to produce adequate mandibular stock.

Distraction can provide even more needed bone in previously deficient areas treated by grafts or flaps if undertaken in appropriate patients. Transport distraction osteogenesis uses the principles of distraction to fill bony defects such as in the calvarial skull or mandible. In addition, techniques such as fat grafting may improve transport distraction osteogenesis by increasing the bone density of the regenerate.

10.10 Conclusion

Distraction osteogenesis is a powerful tool for transforming the craniofacial skeleton through a carefully planned, gradual approach. Advantages include shorter operating times and concurrent expansion of the overlying soft tissue, with fewer complications and less relapse. Distraction does require participation from and communication with families, with acceptance of the longer course of treatment. Given the benefits, the clinical uses are expanding. Technologies such as CAD/CAM and other advances have improved the planning and reproducibility of distraction. Growth modification and overcorrection should be employed when possible. Clinical results and outcomes will continue to be improved with continued study and longitudinal follow-up (► Fig. 10.14).



Fig. 10.14 Distraction osteogenesis represents a powerful tool in craniofacial surgery and can be employed in conjunction with other techniques in a longitudinal fashion to optimize patient outcomes. A patient with Crouzon's syndrome (a) preoperatively, (b) after Le Fort III distraction, and (c) and following a Le Fort I, bilateral sagittal split osteotomies (BSSO), genioplasty, and subsequent rhinoplasty at skeletal maturity.

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11 Orthognathic Correction of Jaw Deformities

Rushil Dang and Cory M. Resnick

Summary

Orthognathic correction of jaw deformities in the pediatric population aims to correct the dental occlusion, along with addressing an equally important goal to improve facial aesthetics. A comprehensive presurgical workup and evaluation must include intra- and extraoral examinations, facial photographs, diagnostic radiographs, mounted dental models, occlusion records, and a facebow transfer. In skeletally immature patients, the nature of the deformity, the operations necessary for correction, and the psychosocial impact of surgery must all be considered when determining the appropriate timing for an operation. Recognition of common facial patterns can be helpful in determining appropriate corrective operations. The most common operations used in orthognathic surgery are the Le Fort I (LFI) osteotomy for repositioning of the maxilla and the bilateral sagittal split ramus osteotomies for movement of the mandible.

Keywords: orthognathic surgery, Le Fort I, bilateral sagittal split osteotomy, ramus osteotomy, facial growth, complications

11.1 Introduction

As many as 5% of adolescents in the United States have a dento-facial deformity that cannot be corrected with orthodontic treatment alone. Advances in operative and anesthetic techniques since von Langenbeck's original description of a maxillary osteotomy in 1895 have allowed maxillary and mandibular osteotomies to become safe and predictable. In the last decade, the combination of three-dimensional treatment planning with low-profile rigid fixation has ushered in a new era of precision in orthognathic surgery.

11.2 Diagnosis and Treatment Planning

Evaluation begins with comprehensive facial and oral examinations, photographs, radiographs, mounted dental models, registration of the occlusion, and a facebow transfer.

The facial examination is performed in natural head position, which is achieved by asking the patient to focus on a distant point at eye level. The shape of the face is characterized, such as square or round, and the facial index (ratio of height to width) is calculated. Normal facial indices are 1.35:1 for men and 1.3:1 for women. The face is evaluated for symmetry at all levels. Vertical facial proportions are assessed by measuring the distances from trichion to glabella, glabella to subnasale, and subnasale to menton; each section should comprise approximately one-third of the total facial height. The lower facial third can be further divided from subnasale to stomion and from stomion to menton, which should account for the one-third and two-thirds, respectively, of the entire lower facial one-third. Transverse proportions are evaluated by partitioning the face vertically

into five equal sections that are each the width of the eye from the medial canthus to the lateral canthus. Shape and symmetry of the nose are determined. Lip morphology and posture are evaluated both in repose and in function. During repose, 2 to 4 mm of the maxillary incisor teeth should be visible. In full smile, the canine tips should be seen without excessive gingival show or dark buccal corridors. Presence and depth of the labio-mental sulcus are evaluated.

On intraoral examination, the presence and health of the complete adult dentition, the dental midlines in relation to the midsagittal plane, the angle classification of occlusion, overjet, and overbite are determined. The occlusal plane is evaluated from both the frontal and the lateral positions. Study casts are used to analyze the intra-arch and interarch relationships.

The radiographic evaluation has historically consisted of a series of plain X-rays, including panoramic, lateral cephalometric, and posterior-anterior cephalometric images. This two-dimensional series has largely been replaced by computed tomography (CT) or cone-beam CT, from which two- and three-dimensional images can be extracted. Three-dimensional imaging has improved accuracy in the planning and simulation of surgical movements (► Fig. 11.1) and in evaluation of relevant anatomic structures, including the airway space, temporomandibular joints, and inferior alveolar nerve canals.

The preoperative assessment also includes an analysis of the patient's level of skeletal maturity. The chronological, skeletal, and dental ages; the nature of the deformity and required corrective operations; and the effect of the deformity on psychosocial development are all considered when determining the appropriate time for an operation. Growth of the maxilla is completed before that of the mandible, with the mandible closely following the skeletal growth pattern. Girls typically reach skeletal maturity between 15 and 16 years of age; boys continue to grow until 16 to 18 years of age or longer. Menarche is a useful marker for the growth cycle for girls, as skeletal growth is usually completed by 2.5 to 3 years thereafter. Evaluation of vertical height, use of hand-wrist radiographs, and overlays of tracings of serial lateral cephalograms can be used to assess the timing of facial growth. An operation involving only advancement of the mandible in the direction of growth can be pursued before skeletal maturity, with low risk for postoperative mandibular growth leading to a new malocclusion. Movements that are not in the direction of growth and bimaxillary surgery should be delayed until after the cessation of growth, unless psychosocial indications dictate earlier operation.

Recognition of common facial patterns can be helpful in determining appropriate corrective operations. Examples include the following:

- Class III skeletal pattern: This is most commonly caused by maxillary hypoplasia but can be due to mandibular hyperplasia or a combination of these. It is corrected by maxillary advancement, mandibular setback, or both (► Fig. 11.2).
- Class II skeletal pattern: This pattern is typically caused by mandibular hypoplasia and is corrected by mandibular advancement (► Fig. 11.3).

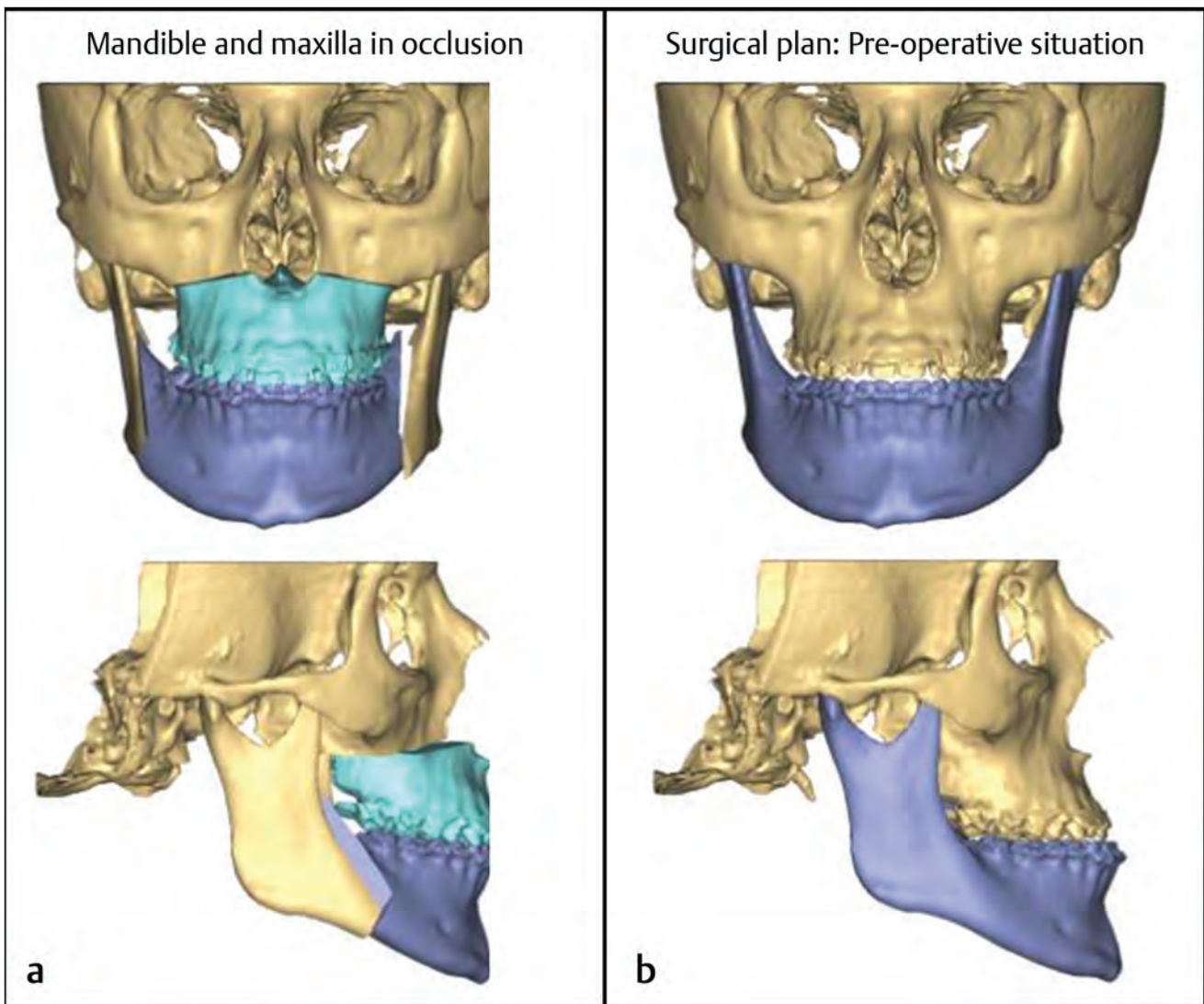


Fig. 11.1 Three-dimensional virtual surgical planning images from a patient with maxillary hypoplasia. (a) Preoperative images. (b) Planned skeletal movements.

- Vertical maxillary excess (VME): It is characterized by excess gingival show, a steep mandibular plane angle, increased total anterior facial height, and decreased contribution of the upper facial height to the total facial height. An anterior open bite is often present. It is corrected by superior repositioning of the maxilla, often with flattening of the occlusal plane. Bimaxillary surgery is often required.
- Facial asymmetry: It presents with various etiologies, including asymmetric mandibular growth, condylar hyperplasia or resorption, trauma, and malformations such as hemifacial microsomia. It is corrected by bimaxillary surgery. The ideal vertical position of the maxilla must be determined in order to identify the proper fulcrum of rotation for the correction.

11.3 Operative Management

The most common operations used in orthognathic surgery are the LFI osteotomy for repositioning of the maxilla and the

bilateral sagittal split ramus osteotomies (BSSO) for movement of the mandible.

11.3.1 Le Fort I Osteotomy

The LFI osteotomy is a versatile procedure that allows the maxilla to be repositioned in three planes of space.

Technique

1. Arrange for the use of controlled hypotensive anesthesia, which reduces bleeding from the rich network of small vessels at mucosal and bone edges that cannot be easily controlled with surgical techniques. The need for transfusion in orthognathic surgery has been nearly eliminated by using this technique.
2. Local anesthesia with vasoconstrictor is administered into the maxillary buccal vestibule.

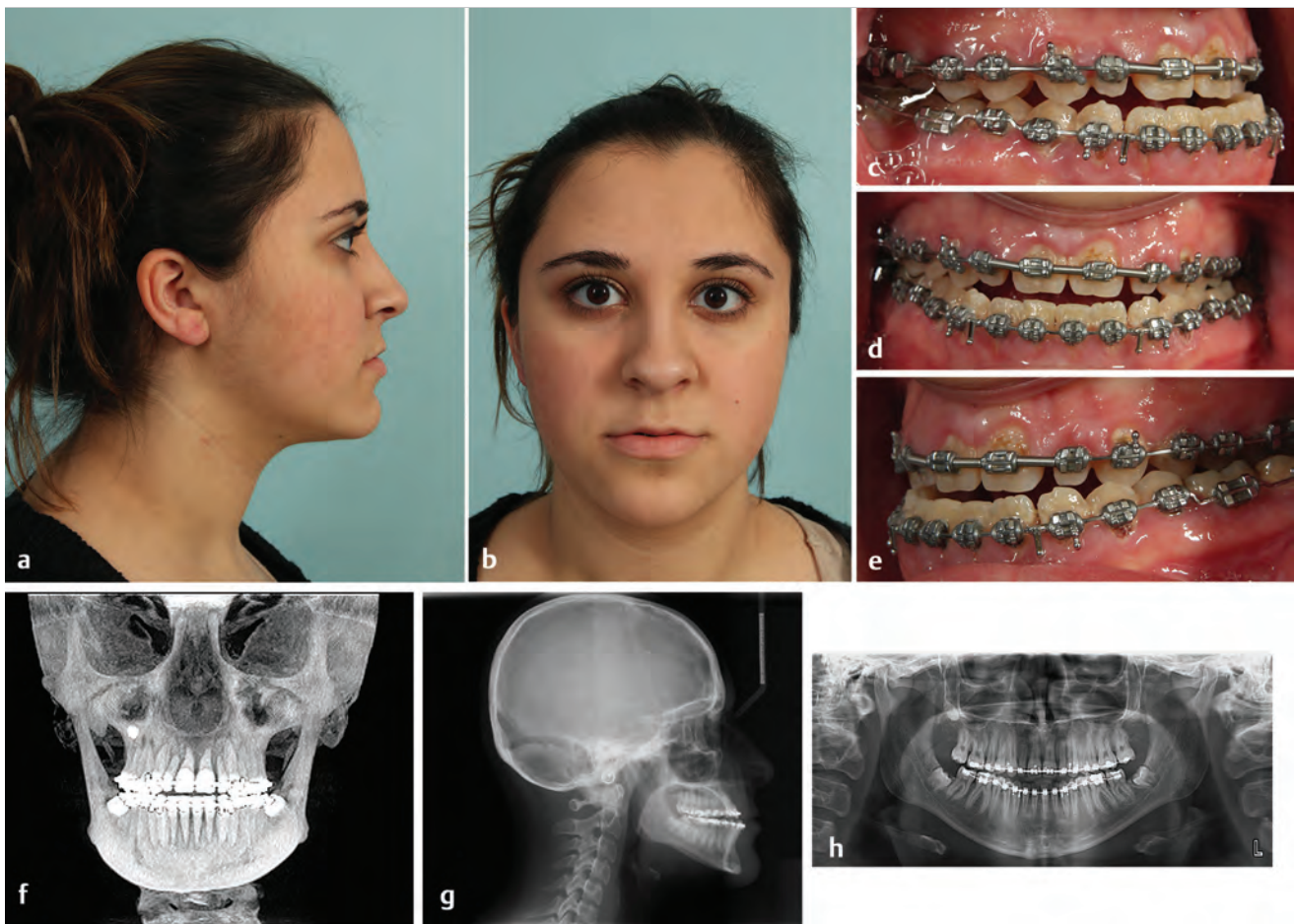


Fig. 11.2 A 20-year-old woman with a class III skeletal pattern and a lower facial asymmetry. (a,b) Preoperative photographs demonstrating a thin and poorly supported upper lip. (c–e) Preoperative dental occlusion. (f–h) Preoperative radiographs showing maxillary sagittal hypoplasia and mandibular asymmetry.

3. An external reference marker is placed at the nasion by using a Kirschner's wire or a bone-fixation screw. Measurements are made from this reference to reproducible midline and lateral maxillary landmarks to facilitate precise repositioning in the vertical plane. Internal reference markers are inadequate.
4. A horizontal incision is made in the gingivobuccal sulcus 5 to 10 mm superior to the mucogingival junction, from first molar to first molar. The submucosal incision is brought inferiorly near the midline to avoid perforation of the nasal mucosa.
5. A subperiosteal dissection is performed along the anterior maxillary walls superiorly to the infraorbital foramina, medially to expose the piriform rims, and laterally to the zygomaticomaxillary sutures. Dissection is extended posterolaterally to the pterygomaxillary fissures.
6. A Freer's or Cottle's elevator is used to dissect the nasal mucosa from the piriform rims and maxillary crest.
7. A reciprocating or piezoelectric saw is used to create a horizontal osteotomy 5 mm superior to the root apices of the canine and first molar teeth, from the zygomaticomaxillary buttresses to the piriform rims on each side.
8. A long curved osteotome is inserted into the pterygomaxillary fissure at a level at and inferior to the horizontal osteotomy on each side. The osteotome is driven in anterior, medial, and inferior directions to separate the posterior maxilla from the pterygoid plates.
9. The osteotomies at the posterior maxillary walls are completed with a small osteotome.
10. A guarded nasal septal osteotome is driven posteriorly and inferiorly along the maxillary crest to separate the caudal nasal septum and vomer from the maxilla.
11. An osteotome is used to fracture the lateral nasal walls to the perpendicular plates of the palatine bone.
12. Downward pressure is applied to the maxilla to create a down fracture. If significant resistance is encountered, the surgeon should stop and reevaluate each of the osteotomies.
13. The descending palatine arteries are identified on each side and evaluated for tears. If necessary, the vessels can be clipped or cauterized, without increasing the risk for vascular compromise of the maxilla.
14. The maxilla is mobilized by stretching the soft tissue envelope by using Rowe's disimpaction forceps and/or pterygoid lifts. Mobilization of the maxilla until it fits passively into the surgical splint in the planned position is imperative.
15. Segmentation of the maxilla can be performed as dictated by the treatment plan.



Fig. 11.2 (Continued) (i–l) Postoperative facial and intraoral photographs after Le Fort I osteotomy, with advancement and anterior disimpaction, and bilateral sagittal split osteotomies, with mandibular rotation. (m–o) Postoperative radiographs demonstrating the planned osteotomies, skeletal movements, and internal fixation.

16. The maxilla is then positioned into the prefabricated surgical splint, and maxillomandibular fixation is applied with wires.
17. The maxillomandibular complex is rotated superiorly while posterior and superior pressure is applied to the mandibular angles to fully seat the condyles. Bony interferences are identified and reduced.
18. Vertical repositioning of the maxilla is determined by observing the relationship between the maxillary central incisors and the upper lip and by measurements from the external reference marker. Measurements to lateral maxillary landmarks are repeated to ensure that no undesired cant of the maxilla has been introduced or that a preexisting cant has been corrected.
19. The nasal septum is evaluated to ensure that it is not buckled. A caudal septoplasty can be performed if necessary.
20. With the maxillomandibular complex rotated to its planned position and the condyles fully seated, fixation is applied using miniplates at the piriform rims and zygomaticomaxillary buttresses.
21. The maxillomandibular fixation is released, and the occlusion is checked. Unplanned occlusal discrepancies must be corrected; failure to recognize and correct a malocclusion at this point will compromise the operative result.
22. The wounds are copiously irrigated.
23. Widening of the alar base is reduced, as desired, with an alar cinch suture.
24. The mucosal wounds are closed with resorbable sutures. The external reference marker is removed.

11.3.2 Sagittal Split Ramus Osteotomies

Sagittal split ramus osteotomies have surpassed vertical ramus osteotomies as the most common mandibular operations in orthognathic surgery because of their versatility in correcting a

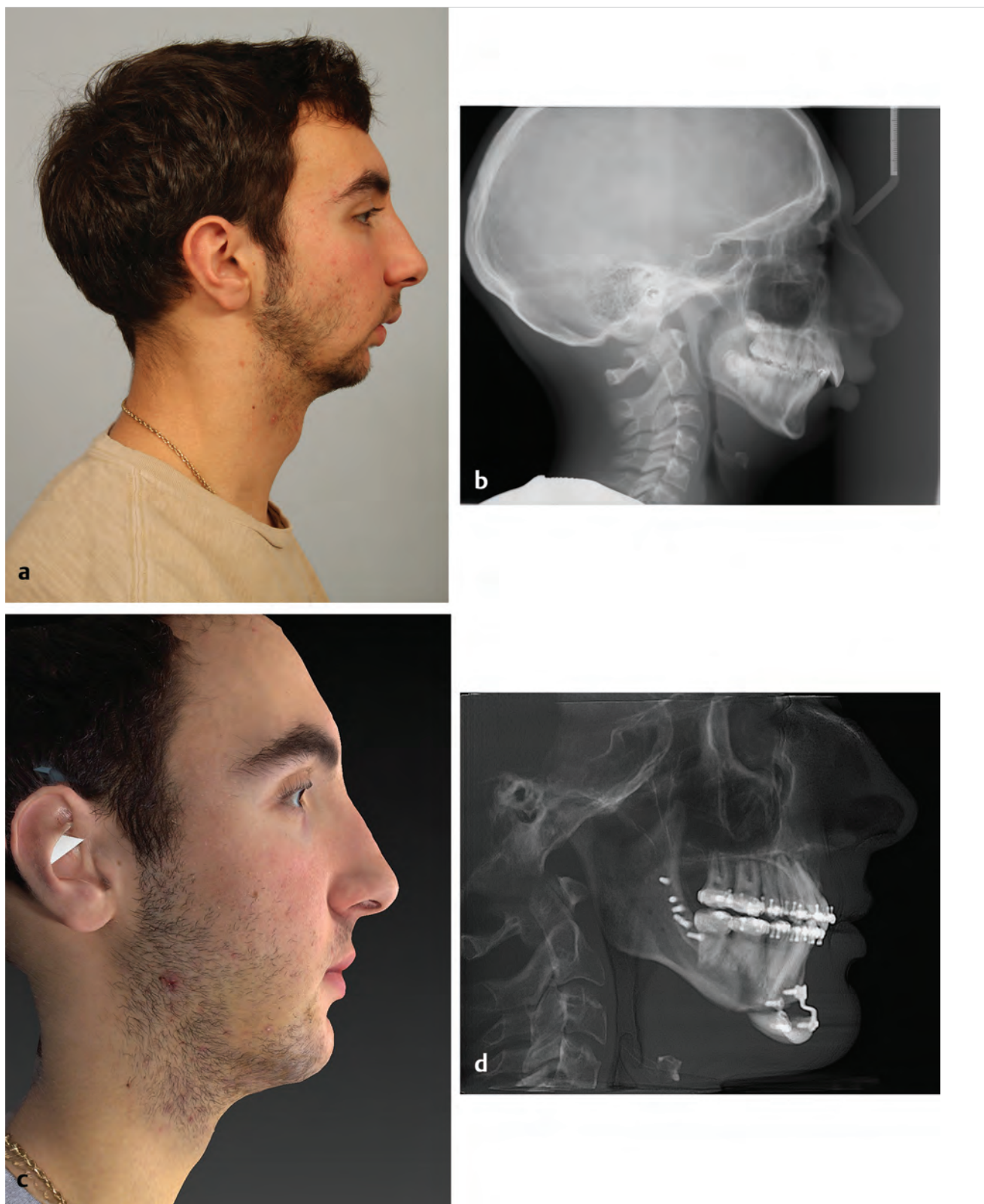


Fig. 11.3 An 18-year-old man with a class II skeletal pattern. (a) Preoperative photograph demonstrating a retrognathic mandible with a convex facial profile. (b) Preoperative lateral cephalometric radiograph showing mandibular sagittal hypoplasia. (c,d) Postoperative three-dimensional photograph and radiograph after bilateral sagittal split osteotomy, with advancement of the mandible and advancing and lengthening genioplasty.

range of mandibular deformities and the ability to routinely apply rigid internal fixation.

Technique

1. Local anesthesia with vasoconstrictor is administered.
2. An incision is made in the buccal vestibule, extending from the external oblique ridge to the area adjacent to the second premolar tooth.
3. The buccal aspect of the mandible is exposed in a subperiosteal plane.
4. Dissection is continued to the inferior border of the mandible. A J-stripper is used to release all muscular attachments from the inferior border in the area of the first molar tooth.
5. Dissection is continued along the anterior ramus to the coronoid process, with stripping of the attachments of the temporalis muscle. A Kocher forceps is applied to the tip of the coronoid process to retract the soft tissues.
6. Dissection is continued on the medial aspect of the mandible, with attention to remain subperiosteal along the natural lingual concavity. The lingula and the mandibular foramen are identified.
7. A Freer's elevator is used to release the attachments of the inferior alveolar neurovascular bundle from the lingula, allowing the bundle to be retracted away from the planned osteotomy site.
8. With the neurovascular bundle protected, a reciprocating or piezoelectric saw is used to create a corticotomy through the lingual mandibular cortex extending from the post-lingula depression to the superior border of the mandible and oriented parallel to the occlusal plane.
9. The cut is continued monocortically along the superior border of the mandible buccal to the dentition to the area of the first molar tooth.
10. At the first molar, the cut is redirected to extend through the buccal cortex toward the inferior border of the mandible.
11. An inferior border osteotomy is then created, extending from the buccal corticotomy and including both buccal and lingual cortices. Special attention is paid to this osteotomy, given that an inadequate cut at the inferior border is a common reason for an undesirable fracture pattern.
12. Osteotomies are used to refine each corticotomy.
13. The mandible is then split using a Smith's spreader and an inferior border spreader. The split is completed slowly while observing for even separation across all bone cuts. Unequal separation is addressed by further refining the cuts in the affected areas before completing the split.
14. After completion of the split, the inferior alveolar neurovascular bundle is identified. The bundle must be completely free from the proximal segment. If a portion of the bundle remains within the proximal segment, a Freer, nerve hook, small osteotome, or piezoelectric saw can be used to free the remaining attachments.
15. The wound is packed, and attention is turned to the contralateral side, where the steps described earlier are repeated.
16. The distal segment of the mandible is positioned into the surgical splint against the maxillary dentition, and maxillomandibular fixation is applied.
17. The proximal segments are repositioned such that the condyles are fully seated in their fossae and the inferior borders of both the proximal and distal segments are aligned.
18. Bony interferences between the proximal and distal segments are reduced as necessary.
19. Fixation is applied on each side by using a minimum of three interposition screws, a plate at the distal osteotomy site, or a combination of these.
20. Maxillomandibular fixation is released, and the occlusion is checked. Unplanned occlusal discrepancies must be corrected; failure to recognize and correct an incorrect occlusal relationship at this point will compromise the operative result.
21. The wounds are copiously irrigated.
22. The mucosal wounds are closed with resorbable sutures.

11.4 Complications of Orthognathic Surgery

11.4.1 Hemorrhage

Since the introduction of hypotensive anesthesia, hemodynamically significant intraoperative bleeding is rare during orthognathic surgery. Major bleeding is most likely to occur during the LFI osteotomy from the descending palatine or sphenopalatine arteries. During the pterygomaxillary disjunction, bleeding can be encountered from the pterygoid plexus of veins or, less likely, from the internal maxillary artery and its branches. Delayed hemorrhage may also occur several weeks after an LFI, most often presenting as severe epistaxis. This is due to bleeding from a pseudoaneurysm that formed as a result of trauma to an intraoperative vessel wall. During sagittal split osteotomies, bleeding most often occurs from the inferior alveolar artery. The facial artery or vein can also be damaged during inferior border osteotomy.

11.4.2 Neurosensory Impairment

Sensory deficits involving branches of the fifth cranial nerve commonly occur after mandibular operations. The inferior alveolar nerve is most often affected, followed by the lingual nerve. Alolayan and Lueng reported neurosensory deficits in 35.4% of patients 6 months after sagittal split osteotomies. Ow and Cheung found a 27.8% rate of persistent neurosensory deficit 1 year after sagittal splits (Ow and Cheung 2009). A transient sensory change of the infraorbital nerve is common after LFI osteotomies, but it typically completely recovers within 6 months.

11.4.3 Malocclusion

Incorrect positioning of the mandibular condyles during fixation or unrecognized bony or occlusal interferences can lead to an early postoperative malocclusion. The most common etiology of an immediate postoperative malocclusion is incomplete seating of the mandibular condyles during repositioning of the proximal segments after sagittal split osteotomies. When maxillomandibular fixation is released, the mandibular condyles move posteriorly to their fully seated positions, resulting in

increased overjet and anterior open bite. Management of an early postoperative malocclusion may require reoperation. Late malocclusion may occur due to orthodontic or skeletal relapse, postoperative growth, unresolved parafunctional habits, or resorptive processes.

11.4.4 Infection

Early postoperative infections occur in 3.1 to 7.4% of patients after orthognathic surgery. Age, medical comorbidities, type and length of the operation, and the use of perioperative antibiotics may affect the risk for infection. Infection rates are increased by concomitant insertion of alloplastic materials such as polyethylene implants.

11.4.5 Fixation Failure

Inadequate fixation and/or early function may lead to hardware failure. Fixation failure most commonly occurs after sagittal split osteotomies and leads to clockwise rotation of the proximal segments and development of an anterior open bite. This is corrected by reoperation with repositioning of the segments and application of additional fixation.

11.4.6 Nonunion

Nonunion is rare and most commonly occurs after an LFI osteotomy. This is typically due to the use of inadequate fixation, early function, or uncontrolled bruxism. Nonunion may present as prolonged pain and fremitus of the maxillary dentition during mastication several months after the operation. Nonunion is managed by reoperation with debridement of fibrous tissue from the previous osteotomy site, bone grafting, application of additional fixation, and strict postoperative non-chewing diet.

11.5 Key Points

- Le Fort I osteotomies and sagittal split ramus osteotomies are versatile procedures for correcting skeletal malocclusions and facial asymmetries.

- Orthognathic surgery requires thorough preoperative evaluation, including facial and oral examinations, photographs, radiographs, mounted dental models, registration of the occlusion, and a facebow transfer.
- Preoperative treatment planning and physical or virtual model surgery are used to create surgical splints that allow accurate reproduction of the plan in the operating room.
- In skeletally immature patients, the nature of the deformity, the operations necessary for correction, and the psychosocial impact of surgery must all be considered when determining the appropriate timing for an operation.
- Accurate and honest assessment of the occlusion in the operating room after release of the intermaxillary fixation is critical to avoid an early postoperative malocclusion.

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Part II

Cleft Lip and Palate

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12 Dental and Orthodontic Management of Cleft Lip and Palate

Richard Bruun and Stephen Shusterman

Summary

Dental and orthodontic management of the patient born with a cleft of the lip and/or palate requires care providers interested, motivated, and dedicated to a lengthy and sometimes complicated process. They should be willing to work in concert with a Cleft Palate Team and well versed in the principles of infant orthopedics (allows a proficient surgeon the best opportunity for primary repair of the cleft lip and nasal deformity) cleft dental anatomy, dental arch coordination, secondary alveolar bone grafting, facial esthetics, and comprehensive orthodontics and dentofacial orthopedics. Patients with clefts require an emphasis on the prevention of dental caries because their teeth are often more susceptible and their burden of care already great. Following the guidelines outlined in this chapter will result in successful treatment for the patient as well as an enjoyable and rewarding experience for the pediatric dentist, orthodontist, surgeon and other team members providing care.

Keywords: teeth, malocclusion, cleft lip and palate, alveolar bone graft, presurgical infant orthopedics, nasopalveolar mold-ing (NAM), latham, pediatric dentist, orthodontics

12.1 Introduction

Infants born with cleft lip and palate should be evaluated by the cleft team, including the pediatric dentist, within days of birth. Pediatric dentists are ideally trained to be a vital part of the cleft team. Their training programs include growth and development, behavioral guidance, guidance of the developing occlusion, preventive dentistry, an introduction to anesthesiology, and operating room protocols. The pediatric dentist has a responsibility to instruct parents in a preventive dental program, to prepare parents for the long dental road ahead, and to actively manage any program in presurgical orthopedic preparation that the cleft team chooses to utilize.

12.2 Dental Care Prior to Bone Graft

Parents of newborns or any young child are responsible for preventive dental care, especially in the case of a child born with cleft lip and palate. Before teeth erupt, the gum pads (alveolar ridges) should be wiped clean after feeding or nursing. By the sixth month of life, the first teeth usually erupt—mandibular teeth erupt before maxillary teeth. It is important to note that there is considerable variability in the timing and pattern and that precocious or delayed eruption should not be cause for great concern or the exposure of radiographs. In children with clefts, the teeth may erupt more slowly; they may be more delayed in the cleft quadrant(s) and slower in males than females. When teeth do erupt, the method of cleaning can be

changed to cleaning with a moist gauze sponge or a clean moist washcloth. As the number of teeth increase and the child begins to walk, the method of tooth cleaning should be changed to brushing, first using nonfluoridated toothpaste and then moving on to fluoridated toothpaste at about 3 years of age, when the child can be instructed not to swallow the often-good-tasting toothpaste. Systemic fluoride should be available through the water supply or via supplementation. The child's pediatrician or dentist should be familiar with the dosing and prescribe appropriately.

During the first year of life, a well-baby visit to the dentist is also recommended by the American Board of Pediatric Dentistry as well as by the American Board of Pediatrics. This initial visit establishes a dental home for the child. It also affords the parents the opportunity to ask questions and to the dentist to educate the parents in preventative methods as well as in dental issues that pertain particularly to children with clefts. It is important to note that even the most minimal instance of clefting may not be without its dental consequences, such as malformed teeth, missing teeth, extra teeth, dysplastic enamel, and even small bony defects.

The early years of childhood should be marked by efforts to preserve the existing teeth and their bony support. Supernumerary palatal teeth, teeth in the cleft, or dysplastic teeth should not be removed, and every effort should be made to keep them caries-free. These teeth are difficult for the child or the parents to brush, thus putting the child at an increased risk of developing dental decay; however, the literature is mixed with regard to the caries rate in children with clefts. In case of carious lesions on malpositioned or malformed teeth, restoration is recommended before the onset of infection. Referral to the pediatric dentist may be indicated, as his or her training in the management of young children will make any necessary treatment as atraumatic as possible.

The issue of the appropriateness of radiographs in the developing child is always a concern for the parents and the cleft team. Cavity-detecting radiographs, exposed no more frequently than every 18 months, are indicated in growing children when posterior teeth contact, when hygiene is less than ideal, and when the risk of caries is high enough to raise the issue of interproximal caries. A child should not be exposed to panoramic or periapical radiographs to determine dental development or to image teeth in the area of the cleft until the child has reached the early mixed-dentition stage (some permanent teeth erupted) or when timing for bone graft is being determined. Just prior to bone graft and to provide the cleft team with more complete imaging, cone beam computed tomography (CBCT) is the radiographic image of choice. With this three-dimensional (3D) reconstruction, proximity to the cleft and the position of teeth can be more accurately visualized. This type of image can also provide panoramic and cephalometric views, by reconstruction. Radiation exposure, given the need for information, is acceptable, and less than that from a medical CT.

12.2.1 Presurgical Infant Orthopedics—Complete Cleft of the Lip and Palate

If the team chooses to go directly to lip repair, it is assumed that the pressure created by the repaired lip will close the alveolar cleft and that pressure on the lip will not affect the esthetic outcome of the surgical scar. However, the authors feel that, in most cases, the repaired lip should not be the sole mediator of pressure on the alveolus. Unavoidable tension on the repaired lip will result in a more prominent surgical scar and the cleft segments will collapse, with the lesser segment falling behind the greater segment in unilateral clefts and both posterior segments remaining behind the premaxilla in bilateral clefts. These failures result in increased complexity of future orthodontic treatment, given later correction of this bony collapse is more difficult. Although the resulting dental crossbites may be corrected, the dentition will lie on a malpositioned bony base. Eventual relapse of the malocclusion is therefore more likely, and lip support, as a prime mediator of facial esthetics, may be compromised.

If the cleft team wishes to incorporate presurgical orthopedics in their treatment protocol, there are several alternatives, which are as follows:

- Taping of the lip +/- the use of a hat or bonnet to provide retraction forces.
- Nasoalveolar molding (NAM) (removable).
- Fixed presurgical orthopedic treatment (FPOT).

Taping of the Lip

Taping of the lip provides another opportunity for pressure on the alveolar segments to retract the more anteriorly placed alveolar segment and close the alveolar cleft before surgical repair. However, placement and retention of the tape is difficult, and replacement of the tape, which must be done frequently, is even more difficult for the parents. This method of closure may be enhanced by the use of the Dynacleft taping system or a bonnet with Velcro attachments for an elastic strap, which will



Fig. 12.1 Bonnet with elastic traction—infant with bilateral complete cleft lip and palate. This method is not only difficult for parents but also uncomfortable for the infant. Again, this method encourages collapse of the segments in a manner similar to the effect of no presurgical treatment.

cross the upper lip for the anticipated and similar effect (► Fig. 12.1). This method is not only difficult for parents but also uncomfortable for the infant. Again, this method encourages collapse of the segments in a manner similar to the effect of no presurgical treatment described earlier.

Nasoalveolar Molding

Nasoalveolar molding is the most widely adopted of the treatment alternatives before lip surgery. In the early 1990s, Grayson et al described a presurgical removable device to move the palatal segments and mold the nasal cartilages. He and his group perceived that the major drawback of previous presurgical devices was their inability to shape the nose. Nasoalveolar molding, therefore, combines an intraoral removable molding appliance with extraoral taping and adds nasal prongs for the purpose of raising the nasal tip and shaping the alar cartilages. The intraoral appliance is adjusted to mold the alveolar segments in both unilateral and bilateral clefts via the addition of self-cure acrylic in the pressure areas and the alternate removal of acrylic on the side toward which movement is desired. When the segments have moved to reduce the alveolar cleft, nasal prongs are added to raise and shape the alar cartilages. Proponents of NAM state that the more difficult surgical goal is the elevation of the ala and the elongation of the columella and that this appliance supports these goals. Technical problems with NAM include hard-to-obtain/unpredictable posterior expansion and poorly directed retraction of the premaxilla in bilateral clefts as well as difficult-to-adjust nasal prongs, resulting in pressure on the ala that may be either inadequate or overzealous in both bilateral and unilateral clefts. Practical problems include the need for a dedicated in-house laboratory and a technician (to achieve the best results) as well as the need for frequent adjustment visits, placing a significant burden of treatment on the family (► Fig. 12.2).

Fixed Presurgical Orthopedic Treatment

The original sketches by Georgiade and Latham emphasized on pressure to rotate and align the palatal shelves while closing the cleft alveolus in unilateral clefts. In bilateral clefts, it was noted that retraction of the premaxilla was not possible without expansion of the palatal shelves, so space had to be developed between the palatal shelves to accept the retracted and aligned premaxilla. The appliances were designed with hinges and activation screws. In case of bilateral clefts, symmetrical expansion was the goal. Latham later added an “auxiliary

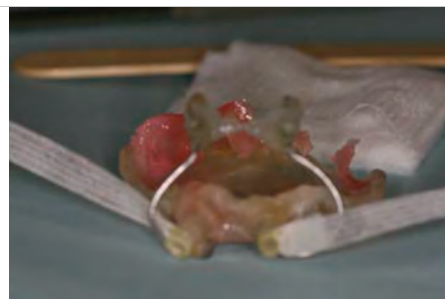


Fig. 12.2 Nasoalveolar molding device.

elastic” to the unilateral design to assist in rotation and closure of the wider clefts. In bilateral clefts, where retraction of the premaxilla was required, a transvomer pin was designed to pass through the bony portion of the anterior vomer just distal to the premaxilla. This pin was attached to the palatal appliance by parallel elastic chains. Both appliances were fixed in place to the palatal shelves with four transpalatal pins.

Our use and studies have supported the routine incorporation of the auxiliary elastic in all unilateral cases. The cleft can be closed by an average of 8.6 mm, thus reducing tension on the repair and enabling the choice of gingivoperiosteoplasty for the surgeon (► Fig. 12.3).

12.2.2 Fixed Presurgical Orthopedic Treatment Protocol of Boston Children’s Hospital

Unilateral Fixed Presurgical Orthopedic Treatment

Infants are examined and parents interviewed between 2 and 8 weeks of age with the goal of lip adhesion or repair at 3.5 months. Diagnostic records are taken, and an impression of the

maxillary dental arch is made. All impressions are taken with the baby lying on the parent’s lap, with feet toward the parent and head on the operator’s knees (knee-to-knee position). Our material of choice is a high-quality alginate with color-changing characteristics. The alginate is placed in a “custom” acrylic tray (made from previous infant impressions and kept in a sterile pouch) (► Fig. 12.4).

Excess alginate is removed posteriorly, before insertion into the mouth. To limit the risk, the tray is not inserted in the mouth until the alginate progresses through several setting stages (purple to pink to white). In our experience, with the baby in this position, the airway has never been an issue, and a detailed impression may be obtained.

Infants with clefts who are to undergo fixed presurgical orthopedics are admitted to the hospital for short stays and brought directly to the operating room suite.

On the day of operation, the infant must have nothing by mouth for 6 hours in advance. An oral endotracheal tube is inserted; the patient is prepped and draped for a sterile procedure, and the oropharynx is packed. The appliance is tried in the mouth to ensure that it fits well; the pins are inserted in slots through the acrylic using a needle holder and set in place by using an orthodontic band seater and a surgical mallet (► Fig. 12.5 and ► Fig. 12.6).

Each pin is approximately 15 mm in length, with a spring loop bend at the head, which will be compressed into the slot in the acrylic to ensure that the pin cannot be lost. When the pins are fully seated and the appliance is in position, the pin slots are covered with a small amount of restorative composite to ensure that the pin will not come out of the appliance, and the composite is set with a curing light. The appliance is activated by rotation of a screw within the cleft site, which extends on a diagonal from the anterior of the lesser segment posteriorly to a receiving cup at the posterior aspect of the greater segment. Initial activation requires 4 to 5 full (360-degree) turns, until the screw is firmly engaged in the cup. The elastic chain extending from the posterior aspect of the greater segment to the anterior aspect of the lesser segment and across to a hook on the greater segment is activated (► Fig. 12.7).



Fig. 12.3 Unilateral fixed presurgical orthopedic treatment device with elastic auxiliary.



Fig. 12.4 (a) Cleft impression tray. (b) Cleft alginate impression.

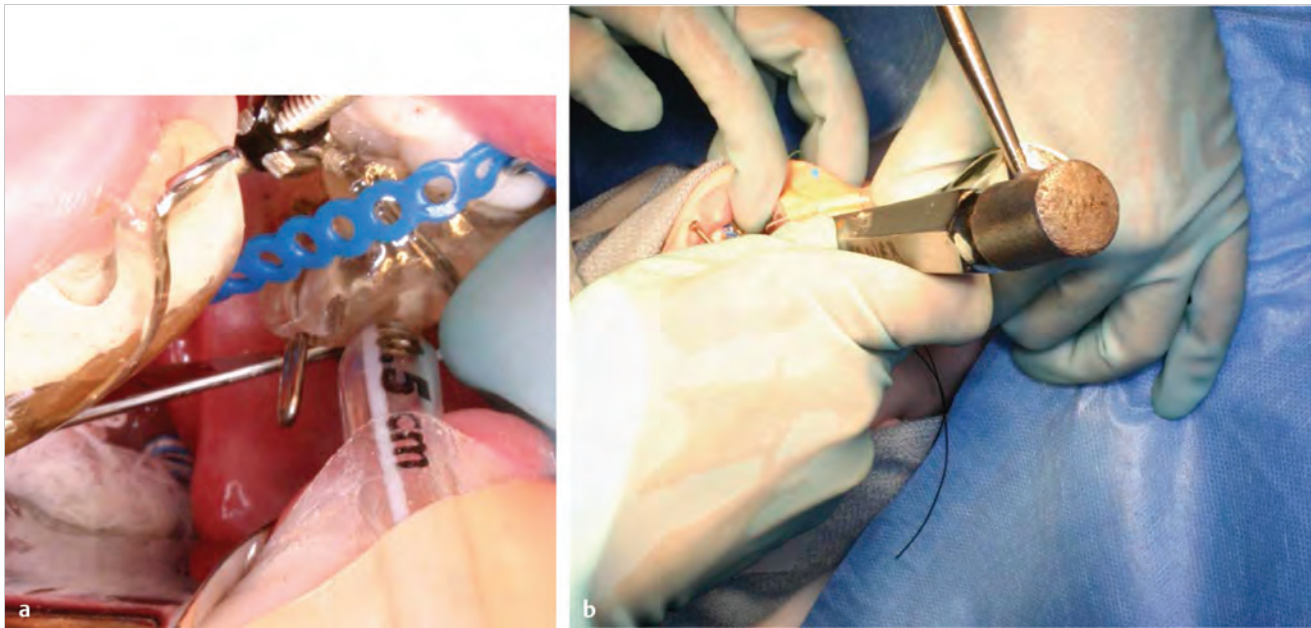


Fig. 12.5 (a) Unilateral fixed presurgical orthopedic treatment during insertion. (b) Mallet and band seater during insertion.

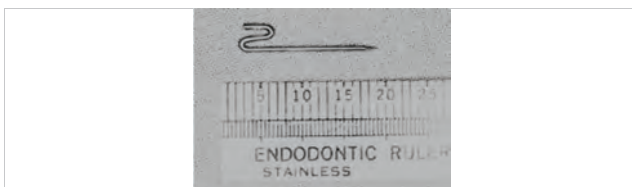


Fig. 12.6 Retention pin.



Fig. 12.8 Bilateral fixed presurgical orthopedic treatment.

The throat pack may then be removed. The child is extubated in the operating room and awakened before being brought to the postoperative care unit. Infants are kept overnight to ensure that they begin to feed orally, that vital signs are stable, and that they are voiding appropriately. Although infants are restless, fussy, and have difficulty feeding immediately postoperatively, a recent study (Bronkhorst et al 2015) has assured us that the classic signs of pain are not present postinsertion.

The infants are then seen at 1 week, 3 weeks, and 5 weeks (more frequently if necessary) to ensure that the appliance is being properly activated at home (the screw is turned) and to permit reactivation of the elastic chain. The appliances are removed at the time of lip repair or lip adhesion.



Fig. 12.7 Unilateral fixed presurgical orthopedic treatment after insertion—chain is activated.

Bilateral Fixed Presurgical Orthopedic Treatment

Infants with bilateral clefts are treated at a slightly older age than those with unilateral clefts (4–8 weeks of age). In the bilateral complete cleft, the anterior margins of the palatal shelves are frequently narrow and the premaxilla on the vomer stalk is markedly anterior and, in some cases, twisted in a lateral or superior/inferior direction. The goal of this treatment is to move the anterior aspect of the palatal shelves apart (transversely), while at the same time retracting the premaxilla to a position between the palatal shelves (► Fig. 12.8).

In the operating room, the junction of the premaxilla and vomer is visualized, and two points are marked along a horizontal line bisecting the cephalad–caudal width of the vomer, approximately 1 and 3 mm posterior to that junction (► Fig. 12.9). Using a handheld chuck and a twist drill (Peeso

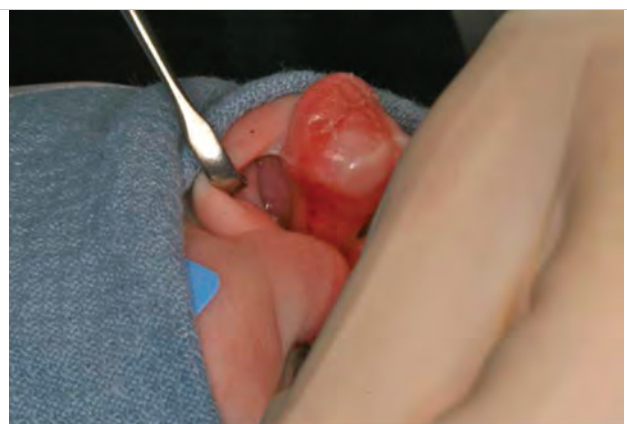


Fig. 12.9 Visualizing junction of the premaxilla and the vomer.



Fig. 12.10 Insertion of U-shaped wire into the vomer.



Fig. 12.11 Elastic chains pulled through bilateral fixed presurgical orthopedic treatment.



Fig. 12.12 Fully seated bilateral fixed presurgical orthopedic treatment—activation screw visualized.

endodontic), two parallel channels are drilled through the vomer at these marks. A U-shaped 0.020 stainless steel wire (8–10 mm in length) is then prepared for insertion through these channels. Elastic chain is then attached to the more distal arm of the “U,” and it is passed through the vomer. Elastic chain is applied to the distal aspect, and the wire bent to prevent its loss (► Fig. 12.10 and ► Fig. 12.11).

The elastic chains are then pulled through the appliance, as it is seated on the palate.

The appliance is secured with four pins in the same manner as the dental maintenance alternative (DMA). The elastic chains are then moved anteriorly and are separately attached with moderate force to the cleats at the anterior edge of each palatal shelf. The activation screw, which is in the posterior midline, is activated (► Fig. 12.12).

The expansion screw in the bilateral appliance (elastomeric chain premaxillary retraction [ECPR]) moves the palatal shelves in a “V” direction and provides enough width to accept the premaxilla. At the same time, the premaxilla is retracted by the elastic chain. The amount of expansion must be closely monitored to prevent a flattening of the nasal tip, excessive width at the alar base, and a prominence of the nasal eminences of the maxillary bone. The patient is monitored at 1 week, 3 weeks,

and 5 weeks to ensure that the screw is activated appropriately and to allow the elastic chains to be reactivated. Feeding is again the main postoperative concern. In addition, mucus and milk curd accumulate between the palatal shelves and are cleaned with suction at each visit.

The impact of presurgical treatment on the family should not be minimized.

Parents of infants born with clefts of the lip and palate have already been significantly challenged in the postpartum stage. Although most parents now know through prenatal ultrasound that their child will be born with a cleft, the immediate mournful reaction after birth (when the reality is confronted) may be overwhelming. Relatives and friends who may not be fully prepared may become part of the problem. One of the advantages of presurgical care is the introduction and support of the cleft team and the knowledge that they are available to help at any time. From the initial impression on, this is a difficult period for parents. Our recent review of the postoperative course of infants who have had Latham-type appliances has shown that pain, as measured by vital signs and Face, Legs, Activity, Cry, and Consolability (FLACC) scores, is not a significant problem. However, it is not our intention to minimize the disruptions of these appliances, which include absence from work, loss of sleep, difficulties

in feeding, the impact on other children in the family, and many more. These are significant disruptions and require that members of the cleft team, including the dentist and the feeding specialist, are available to the parents. However, the result of well-planned and executed presurgical infant orthopedics will leave the child with a dentition that is easily maintained, that has acceptable occlusion and appearance, and that is ready for the next phase of treatment in the mixed dentition.

12.3 Orthodontic Management—Mixed Dentition to the Completion of Treatment

12.3.1 Diagnosis

The diagnostic materials required for the evaluation of the malocclusion associated with cleft of the lip and or palate vary considerably according to the patients' stage of development and the exact diagnosis. Models of the teeth; intraoral and extraoral photographs; and panoramic, lateral cephalometric, and frontal cephalometric radiographs comprise the diagnostic materials required, usually referred to as orthodontic records.

Lateral cephalometric analysis of the patient in *natural head position* (NHP) will provide the orthodontist and surgeons with excellent insight into the nature and magnitude of the skeletal and dental malocclusion (in the sagittal plane) as well as help them understand the perceived appearance of the dentition and the profile. The use of “true” vertical and horizontal reference planes, based on NHP, will help you make the best

decisions regarding not only orthodontic therapy but also the need for, and desired results of, orthognathic surgery (► Fig. 12.13).

In patients with any obvious or suspected bony alveolar defect, it is important to fully evaluate the cleft area or areas before (and following) secondary alveolar bone grafting. This will help determine if there is a bony defect that requires grafting as well as the nature and extent of any such defect. It is also important to determine the usefulness of the teeth in the cleft region and to identify any supernumerary, ectopic, and/or malformed teeth. After the graft, one needs to understand the viability of the new bone in terms of orthodontic tooth movement and as a possible site for one or more osseointegrated dental implants. These tasks are best accomplished using 3D imaging, preferably low-dose CBCT, which provides more information than 2D radiographs and can be used to construct the 2D images that the orthodontist requires. The orthodontist and/or surgeon must become proficient with one or more 3D viewing software programs, such as Dolphin 3-D, in order to fully utilize the information that can be derived from such a study (► Fig. 12.14).

12.3.2 Management

Restoration of the dentition and treatment of malocclusions for patients with cleft lip and palate require particular attention to a number of issues specific to this group as well as an understanding of the overall nature of the treatments necessary, whether they are dental, surgical, or other. It is imperative that the orthodontist acts as a part of the cleft lip and palate team. A coordinated and thoughtfully worked out, and executed, treatment plan will ensure excellent care.

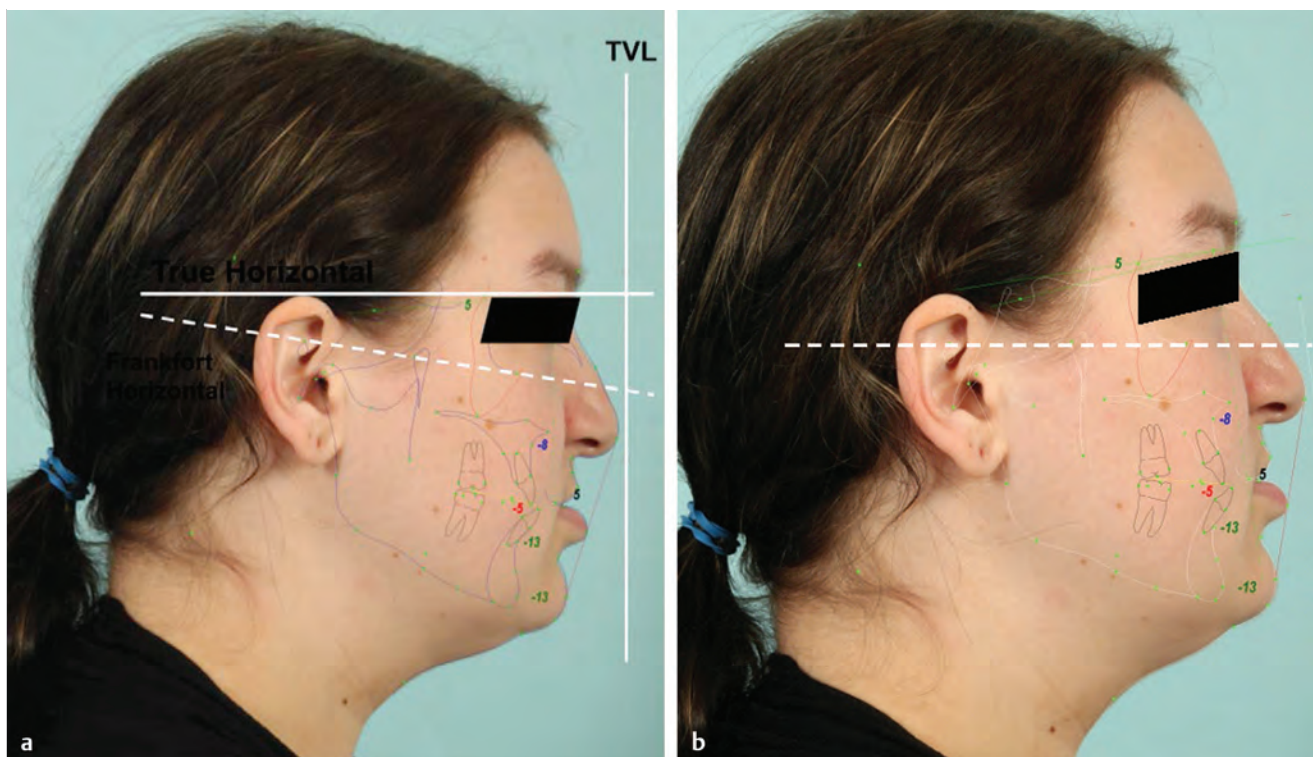


Fig. 12.13 (a) A patient with unilateral complete cleft lip and palate in “natural head position.” (b) Same patient oriented to Frankfort “horizontal” (skeletal).

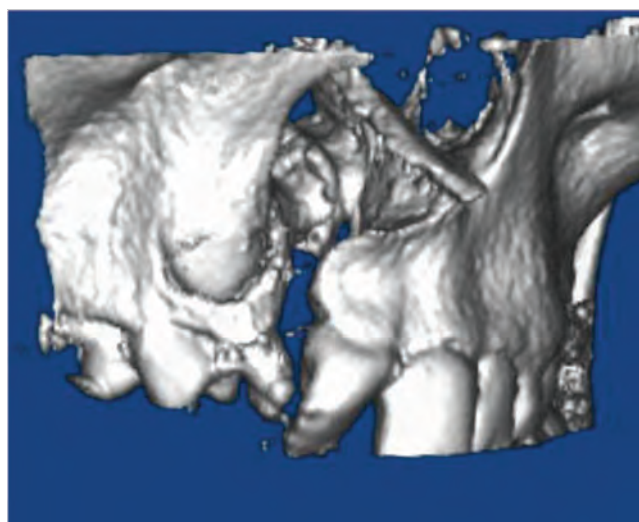


Fig. 12.14 Cone beam computed tomography of right unilateral alveolar defect.



Fig. 12.16 Typical clinical presentation—left unilateral complete cleft lip and palate, early mixed dentition.

In case of any cleft involving the alveolus where there is a bony defect requiring surgical repair (most commonly complete cleft of the lip/palate but also including cleft lip/alveolus with intact secondary palate), cleft orthodontic treatment may be divided into three phases. Phase I is the one that precedes the secondary alveolar bone graft. Phase II is the phase that immediately, or shortly thereafter, follows the graft. Phase III provides definitive or end-stage orthodontia and is coordinated with final restoration (prosthetic reconstruction) of the dentition and orthognathic surgery, when indicated.

12.4 Cleft(s) Involving the Alveolus

12.4.1 Phase I Orthodontics

Most patients with complete cleft lip and palate at this stage present with a significant bony alveolar defect and a constriction of the maxillary dentoalveolar arch, in which we see the

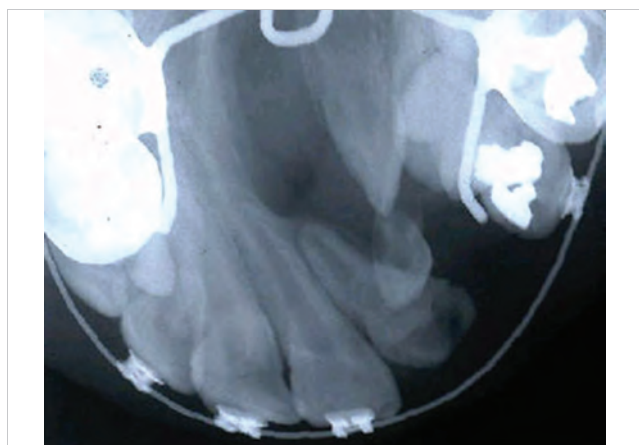


Fig. 12.15 Maxillary occlusal radiograph of left alveolar bone defect.

segment(s) collapsed more severely anteriorly than posteriorly (► Fig. 12.15). In unilateral clefts, there is often more collapse on the cleft side than on the noncleft side. There may be tooth anomalies such as agenesis and dysplasia of a tooth or teeth (most commonly a missing lateral incisor and a dysplastic central incisor adjacent to the cleft), supernumerary teeth, ectopic teeth, and/or malformed teeth. There is often an anterior crossbite reflecting a developing maxillary retrognathism and palatal tipping and/or rotation of the maxillary incisors (► Fig. 12.16 and ► Fig. 12.17). In bilateral complete clefts, the entire premaxilla may be malposed. It is not rare to find a patent or blocked oral–nasal fistula still present at this time.

The goals of phase I orthodontic treatment are to position the dentoalveolar segments so as to approach ideal maxillary arch form and width, to provide the surgeon with a better access to the graft site, and to begin preparation of the occlusion for definitive treatment. It most often takes place in the mid to late mixed dentition, usually between 9 and 11 years of age. Most surgeons perform the alveolar bone graft when the cleft-side canine(s) has formed between half and two-thirds of its root. Orthodontic treatment in the mixed dentition should be started early enough to ensure that maxillary expansion, alignment, and retention are completed by this time. Eruption of the canine into the grafted bone will help ensure viability of the bone graft, which will, in turn, provide adequate bony support for the adjacent teeth and allow for later phases of orthodontic treatment to be successfully performed. It should be noted that, occasionally, a tooth other than the canine might dictate graft timing, following the same principle as the canine would.

Recognizing that the maxillary constriction found in a patient with cleft is usually asymmetric implies that maxillary expansion should likewise be asymmetric. The ideal device to accomplish this is the one that expands the maxilla differentially (anterior greater than posterior) in a concise and predictable manner. In case of unilateral complete clefts, expansion before grafting allows for greater expansion of the lesser segment, resulting in a more symmetric maxillary arch, more closely approximating the form of the mandibular arch. At Boston Children's Hospital, we employ the use of a “fan”-type expansion screw in a fixed maxillary expansion appliance, the *differential MEA* (► Fig. 12.18 and ► Fig. 12.19).



Fig. 12.17 Panoramic radiograph—right unilateral cleft lip and alveolus.

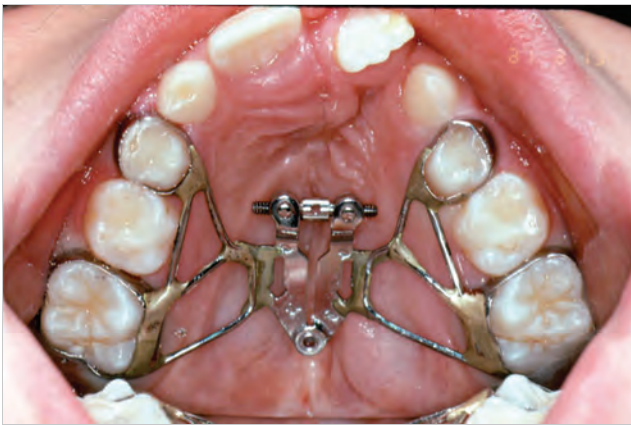


Fig. 12.18 Differential maxillary expansion appliance—preactivation.

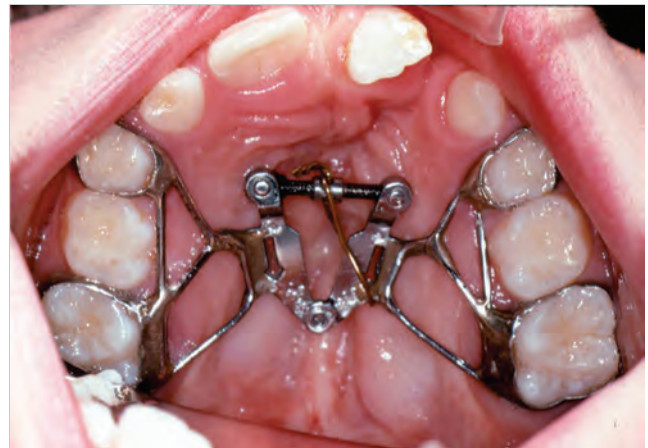


Fig. 12.19 Differential maxillary expansion appliance—postactivation.

The abutment teeth for this appliance are usually the first permanent molars and the first primary molars; however, other teeth in similar relative positions may be used if desired or necessary. By varying the position of the fulcrum (hinge) of the MEA, the ratio of anterior versus posterior expansion can be reliably programmed, so that maxillary expansion results in the proper maxillary arch form and width. In this manner, overexpansion of the permanent molars that might result from the use of a symmetrical expansion appliance is unlikely to occur. The multiple adjustments necessary to manage other often-used expansion devices, such as the “quad-helix” or the “Porter arch,” are also not an issue. Following expansion, the appliance is removed and replaced with a fixed transpalatal arch, with palatal arms extending anteriorly, which should remain in place through the time of the bone graft (► Fig. 12.20).

The magnitude of the expansion desired requires careful analysis of the diagnostic records and an understanding of the likely growth pattern of the patient. A significant number of patients with complete cleft lip and palate become candidates for surgical maxillary advancement at the end of their growth. This is often indicated to provide the best facial aesthetics possible in light of their maxillary deficiency as well as to minimize dental compensations that would otherwise be necessary to achieve a satisfactory occlusion. A common error made by inexperienced orthodontists is overexpansion in phase I, done to



Fig. 12.20 Transpalatal arch—postexpansion retention device.

widen the maxillary arch to correct apparent constriction, while failing to account for the future advancement of the maxillary dental arch.

Orthodontic movement of anterior teeth before bone grafting may be considered when strong indications exist, such as the relief of traumatic occlusion and providing the surgeon with

access to the graft site, and only when adequate bone between root and defect is confirmed by using 3D imaging. Great care should be taken as the root of any incisor adjacent to a bony defect is often separated, on some portion of its root from the defect by very little or no bone. The distopalatal aspect of cleft-side central incisor should be examined carefully on CBCT by reorienting the study along its long axis and viewing it in all three planes of space.

Orthodontic tooth movement of the incisor or incisors in closest approximation to the cleft may lead to perforation of a significant portion of the root into the defect. If this occurs, it will likely lead to compromise of the graft and/or loss of the tooth.

The application of an orthopedic force by means of a reverse-pull headgear (facemask) may be considered as part of this phase of treatment. Several protocols for its use have been advocated and may be considered in cases of mild to moderate maxillary deficiency. This treatment can often be combined with maxillary expansion and should always be done with care being exercised not to overly compensate the dentition (excessively tip the maxillary incisors forward). Increased burden of care, lack of cooperation, and uncertainty of the outcome are the factors that discourage the use of these appliances in some patients (► Fig. 12.21).

For patients with bilateral complete cleft lip and palate, a premaxillary osteotomy may be required in order to better position a particularly malposed premaxilla that cannot be corrected orthodontically (as is sometimes possible by using orthodontic forces on a mobile premaxilla). The premaxilla may be severely vertically malposed (usually inferiorly) and/or rotated in the sagittal plane (anterior nasal spine forward and the incisal edges of the incisor teeth backward).

Less frequently, it may be severely deviated to one side or canted in the coronal plane. The premaxillary osteotomy can be done at the same time as the bone grafts, and care should be

taken by the surgeon so as not to position the premaxilla as superiorly as may seem indicated given that it will usually fail to grow vertically to the same degree as the remainder of the maxilla. The use of both acrylic and orthodontic wire splints will help ensure the stability of the premaxillary segment and the success of the bone graft.

12.4.2 Phase II Orthodontics

The purpose of phase II is to utilize, maintain, and support development of the grafted bone by means of introducing the mechanical stimulation provided by erupting and/or functioning permanent teeth into the graft. This is done by guiding eruption of the canine(s) into the region and/or orthodontically moving teeth adjacent to the site into the newly placed bone, thus encouraging the lengthening and thickening of the alveolar bone and preventing the thinning and shortening (atrophy) that naturally occur over time in any edentulous area. Orthodontic treatment in this phase should also strive to further develop good maxillary arch form and width, align teeth, improve esthetics, and prepare for and simplify definitive treatment at a later stage.

The orthodontist must understand that the natural course of jaw growth in patients with complete (involving the primary and secondary palate) cleft is such that maxillary growth has likely been and will continue to be deficient, resulting in an increasing relative horizontal retrusion of the maxilla to the mandible and to the upper face. As the patient matures, any underbite or open-bite discrepancies between the maxilla and the mandible are likely to worsen, as the mandible grows but the maxilla does not (► Fig. 12.22).

For patients with cleft of the lip and alveolus not involving the secondary palate, maxillary growth has likely been and will continue to be more favorable than in patients with complete cleft. This understanding should be factored into decisions

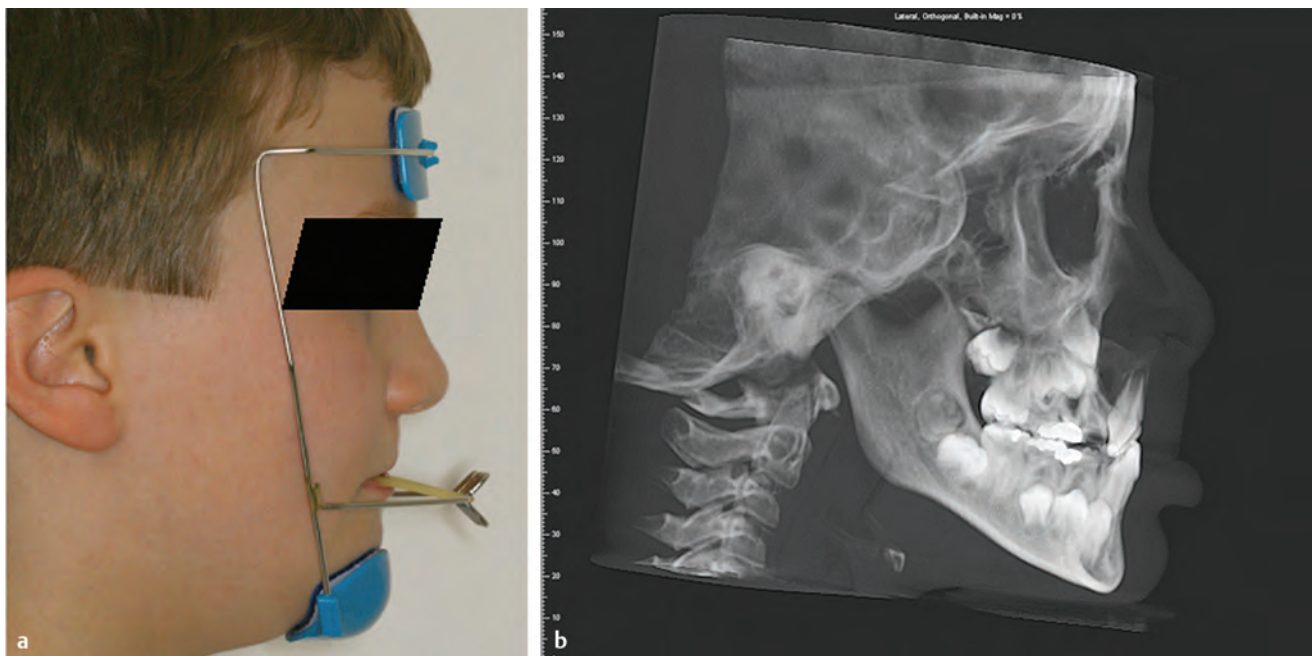


Fig. 12.21 (a) Reverse-pull headgear. (b) Lateral cephalometric radiograph—bilateral complete cleft lip and palate before premaxillary osteotomy.



Fig. 12.22 Unfavorable facial growth in a patient with unilateral complete cleft lip and palate.

about which patients might be amenable to nonsurgical treatment and about the use of orthopedic forces during phase II.

For patients with a substantially unfavorable relationship between the maxilla and the mandible (skeletal class III malocclusion) at this stage, the knowledge that remaining growth will only worsen the problem signals that surgical orthodontic therapy will be indicated in order to provide a satisfactory occlusion and an esthetic profile. With that in mind, the orthodontist should try to minimize the amount and length of phase II, often limiting the treatment to the maxillary arch, whenever possible.

For patients with a favorable or marginally unfavorable relationship between the maxilla and the mandible, the orthodontist may proceed with more substantial treatment during phase II. However, before the completion of growth, it is quite difficult to predict with certainty that a class III malocclusion would not develop. Even a small change (as little as 1–2 mm) will likely require orthodontic retreatment. More significant change could require transition to surgical orthodontic treatment. Caution should be taken to prevent acting on the natural desire that the orthodontist has to use his or her skills to treat a malocclusion that can be better treated in coordination with orthognathic surgery with braces alone. Of particular concern is the marginally class III patients who start phase II orthodontic treatment with the hope that orthognathic surgery would not be required and then begin to show an unfavorable growth pattern (pubertal growth spurt). At this point, the orthodontist should refrain from further compensation of the teeth, given that continuing to treat in this way, combined with continued unfavorable growth, will leave the patient with a poor occlusion, severely compensated teeth, and a skeletal discrepancy between the jaws greatly exceeds the dental discrepancy. Teeth will then need to be decompensated before orthognathic surgery. This “round tripping” of teeth may lead to blunting of roots or even loss of teeth.

A benefit of phase II treatment is the fact that it often takes place during the time at which many other children are

undergoing orthodontics. This allows the patient with a cleft to “fit in” with their contemporaries and reduces the feeling of being different. In addition, the emotional value of improved appearance, even if only a modest change during this phase, should not be underestimated.

Phase II treatment is another good time to consider the possible benefits of treatment involving dentofacial orthopedics for the patient with a cleft who has a developing class III malocclusion that is not excessive in nature. If the patient and family are willing to accept a compromise in the facial esthetics (profile), dentofacial orthopedic treatment may prove useful in mitigating the developing class III malocclusion by encouraging forward maxillary growth and/or discouraging forward mandibular growth. The efficacy of such treatment remains an area of significant controversy within the orthodontic community, as many believe the majority of the changes to be dentoalveolar in nature and therefore the risk of excessive dental compensation (tipping of teeth to camouflage the skeletal malocclusion) to be too high. If the patient’s family wants their child to have a profile that approaches or reaches normal for their ethnic group, dentofacial orthopedics is not indicated except in the mildest manifestations of class III skeletal malocclusions. An alternative to traditional dentofacial orthopedics is the use of so-called “Bollard” plates, which are attached to the maxilla and mandible directly, in the mandibular canine and maxillary malar process regions, respectively, allowing the patient to wear elastics without directly affecting tooth positions but, possibly, affecting maxillary position as far superiorly as the zygomatic arch (► Fig. 12.23).

12.4.3 Phase III Orthodontics

The goals of definitive orthodontic treatment(s) are to provide a stable, healthy, functional, and aesthetic dentition and occlusion as well as to ensure good facial aesthetics. This phase of treatment requires careful coordination with the team and involves orthodontics, with or without orthognathic surgery,



Fig. 12.23 Bollard plates with elastics in place. (This image is provided courtesy of Heymann et al 2010.)

prosthodontics, and periodontics. It must also take into account the facial changes that are likely to result from final lip or nasal revision.

It is best to begin phase III with a decision having been made as to whether or not orthognathic surgery is to be a part of treatment. To proceed, the patient must be skeletally mature enough, so that the team can be confident that there will be little enough further change in the maxillomandibular relationship to make such a choice.

Nonsurgical Orthodontics

The decision to provide definitive orthodontia without orthognathic surgery does not mean that the orthodontist no longer needs to communicate with and coordinate treatment with the cleft team. Understanding the nature and expected outcome of final lip revision and how that procedure will affect tooth show, at rest, during speech, and when smiling should be accounted for by the orthodontist. Furthermore, any dental implants into the grafted bone should be prepared for, as required by the cleft team.

Surgical Orthodontics

An understanding of the planned operation is essential for the orthodontist to prepare the patient properly for orthognathic surgery. Vertical changes, often involving anterior maxillary disimpaction and alteration in the occlusal plane, need to be accounted for during orthodontic preparation, in order to achieve the best possible outcome. Of particular concern is achieving adequate forward incisor angulation and adequate anterior arch width (intercanine, except really interpremolar in this case) when the plan calls for canine substitution of missing or extracted lateral incisor(s).

Surgical orthodontic treatment should begin no sooner than the estimated time that the orthodontist will take to prepare the patient for the operation. In addition, medical history, hand-wrist bone age, and/or serial cephalometric radiographs must ensure completion of skeletal growth by the time of surgery (► Fig. 12.24).



Fig. 12.24 H-W film.

An ideal surgical orthodontic treatment plan will result in an occlusal discrepancy equal to the amount of skeletal movement required to achieve the desired soft tissue profile changes (► Fig. 12.25).

Frequent updates from the orthodontist and study models, photos, and, less frequently, lateral cephalometric radiographs, will assure the surgeon that the treatment is progressing as planned and will produce a more accurate result. Following the operation, the orthodontist continues treatment, using the orthodontic appliances, to help provide stability in the immediate postsurgical period and then to detail the occlusion before removing the braces.

12.4.4 Retention

Retention is important in every orthodontic case, but it is a subject of great concern in the treatment of patients with cleft lip and palate, not only after definitive treatment but also after each phase along the way. Maxillary expansion may be especially subject to relapse in patients with complete clefts of the lip and palate, where transverse dimensional stability must be supported for life due to the lack of a true secondary palate. The use of a fixed transpalatal arch with anterior extensions certainly helps maintain the expansion achieved during phases I and II, but after final treatment, the diligent use of a removable retainer (part-time) is more practical.

12.5 Isolated Cleft of the Palate

Patients with the diagnoses of an isolated cleft palate (ICP) or its associated subgroup, nonsyndromic Robin's sequence (RS), have a markedly different malocclusion than patients with cleft of the lip and palate. This malocclusion is characterized by bimaxillary skeletal retrusion, which is more pronounced in the mandible in case of patients with RS.

12.5.1 Early Treatment (Mixed Dentition)

Early management of the malocclusion present in patients with ICP and RS focuses on the dental crowding, most often found to



Fig. 12.25 (a) A patient with left unilateral complete cleft lip and palate prepared for Le Fort I maxillary osteotomy. (b) Same patient with left unilateral complete cleft lip and palate post Le Fort I maxillary osteotomy.

be present, and the relative mandibular retrognathia seen in RS. Crowding may be severe enough to warrant a “serial extraction” protocol, whereby a series of primary and then secondary (permanent) teeth are extracted at selected intervals in order to minimize the amount of orthodontic treatment. The use of a “functional” appliance to encourage mandibular growth may be considered when a class II skeletal and dental malocclusion exists. The lack of predictable results and the risk of excessive dental compensation are potential complications that should be considered.

12.5.2 Definitive Treatment

When the retrusion of the maxilla and mandible are similar enough in magnitude to bring the relative anteroposterior (AP) malocclusion into a range treatable by orthodontics alone, the treatment may follow a typical course of orthodontia. However, if both jaws are significantly retrognathic, the child may be

predisposed to sleep apnea and/or may be dissatisfied with his or her appearance. If one or both of these issues exist, bimaxillary orthognathic surgery to advance both jaws may be indicated. If one jaw is more retrusive than the other, it may be reasonable to advance only that jaw, given that the other is only mildly retrusive and no sleep apnea exists. If both jaws are only mildly retrognathic, traditional orthodontic therapy should be all that is required.

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13 Unilateral Cleft Lip

Ananth S. Murthy and Paul Durand

Summary

Careful preoperative preparation of an infant with a cleft lip requires addressing feeding issues and ensuring that there are no comorbidities. If a complete cleft lip is present, preoperative dentofacial orthopedics will aid in preferred arch and alar base alignment. A lip adhesion may be pursued to address the arch and alar base alignment, decrease the tension at the repair site, and increase length of the medial element. Meticulous measurements and markings for a rotation–advancement repair allows for accurate placement of the nasal sill, philtrum, Cupid's bow, vermillion, and mucosa of the upper lip. Synchronous nasal correction of the alar cartilage, alar base, caudal septum, as well as the vestibular web is imperative for removing the stigma associated with a cleft.

Keywords: cleft lip, cleft repair, unilateral cleft lip, lip adhesion

13.1 Introduction

The commonly quoted incidence of cleft lip and/or palate of 1 in 1,000 live births does not reveal the complexity of treatment of this congenital anomaly. As a result of a genetic or environmental mishap, the lip, alveolus, and palate may fail to fuse and result in a cleft lip and palate. This cleft may cause problems with breathing, chewing, sucking, speaking, hearing, swallowing, and appearance. It takes a combined effort of teams of health care professionals to undo the effects of this deformity.

Cleft lip represents a failure of the medial nasal process to fuse with the maxillary processes on each side. Several genes are activated and deactivated in the first 3 to 8 weeks of life, which are critical to formation of the lip and nose. Genetic variability of this process of migration results in phenotypic heterogeneity. Consequently, clefts of the lip may be unilateral or bilateral and complete or incomplete. A complete cleft lip may have a skin bridge between the medial edges of the nasal floor, known as a “Simonart's band.” A minor form of an incomplete cleft is referred to as a microform cleft lip (► Fig. 13.1 and ► Fig. 13.2).

It is important to realize that the origin of the cleft can also produce other congenital anomalies. In fact, 35% of all infants with cleft lip and palate have additional associated anomalies. This incidence rises to 54% for infants with a cleft palate alone.

Therefore, when evaluating a patient with cleft lip and palate, it is necessary to perform complete physical and genetic workup.

Periodically, such anomalies occur together and are named after their descriptor, given there is no apparent singular cause. Presumably, the multiple anomalies that make up the syndrome are related. Numerous syndromes that involve orofacial clefting have been delineated. As research in genetics grows, the source of some of these syndromes has been isolated. Common syndromes associated with cleft lip include Shprintzen's (velocardiofacial syndrome; 22q11 microdeletion) and van der Woude's (*IRF6* mutation) syndromes.

With the increasing sophistication of ultrasonography, the anatomy of the lip can now be discerned prenatally. If a complete cleft is present, it may be detected by 16 weeks, and an incomplete cleft can be detected by 27 weeks. A prenatal consultation with the mother can help allay her fears and offer expectations for surgical treatment. Obstetric anxiety can also be reduced with a simple statement of hope, describing accomplishments of other children with cleft lip and palate.

Once a child is born with a cleft, the most pressing concern is feeding. Since such infants are unable to form a seal around the nipple to produce adequate negative pressure required for sucking, they are faced with an immediate problem. This can be ameliorated by using a modified nipple (wider aperture and squeeze chamber), such as a Mead Johnson's or Haberman's nipple, as well as several others that are commercially available. Our team uses Dr. Brown's nipple as a first-line measure, which relies on alveolar compression to open the aperture. This allows the infant to control the flow to some extent.

After the baby is able to feed and demonstrates growth, surgical correction can be pursued. Although not supported by scientific evidence, Wilhelmsen and Musgrave emphasized the “rule of 10”: weight 10 lb, hemoglobin 10 grams, and age 10 weeks. However, in some cases, the option of a lip adhesion may be considered before definitive lip and nasal correction. If the lip adhesion is performed after 10 weeks, a definitive repair may be scheduled 2 to 3 months later.

13.2 Evolution of Care

The first cleft lip repair was reportedly performed by an unidentified Chinese physician in 390 AD. It was not until 1310 when Jehan Yperman (1295–1351), a Flemish surgeon,

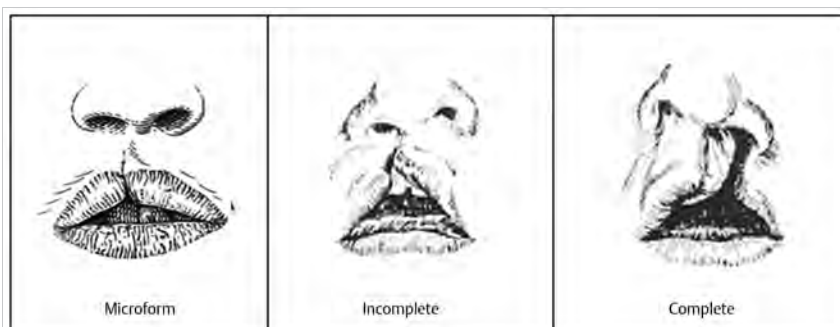


Fig. 13.1 Classification of unilateral cleft lip.



Fig. 13.2 Case example: Note the presence of a “Simonart’s band.” In such cases, the alveolus is often well aligned and so is the alar base. Therefore, active dentofacial orthopedics and lip adhesion were avoided, and she underwent a one-stage lip and nasal correction. At 3 years of age, the girl’s nasal correction is held and will not require any further revisions.

provided the first description of the procedure. In his repair, Yperman recommended a straight-line closure reinforced by passing a long needle through each side of the lip and fixing the needle shaft with a figure-of-eight suture. This was refined by French surgeon Ambroise Pare, who, in 1575, provided the first illustration of the lip correction with a similar technique. Early techniques in cleft lip repair mostly involved a straight-line closure. Later, these evolved to include other methods using geometric advancement flaps and Z-plasties. By the mid-20th century, the two most widely used repairs were those of LeMesurier and Tennison.

While stationed as a Navy surgeon during the Korean War, Dr. Ralph Millard grew dissatisfied with the results he obtained from the LeMesurier technique. He noted that although most of the Cupid’s bow, one philtral column, and the philtral dimple

were all intact on the medial side of the cleft, they were all rotated upward. Rather than disturbing these structures as in previous repairs, he believed that these only need to be released from their attachments and rotated downward to achieve normal position. These observations led Millard to develop the rotation-advancement technique, which to this day remains the basic foundation for unilateral cleft lip repair in most cleft centers throughout the world.

13.3 Normal and Cleft Lip Anatomy

Successful correction of a cleft lip is directly related to the ability to restore the appearance and functionality of the upper lip,

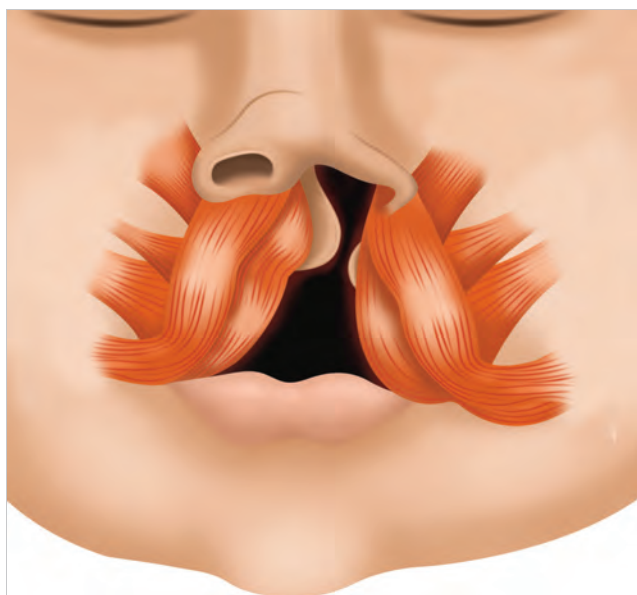


Fig. 13.3 Muscle bundles are redirected due to the cleft lip. The medial portion of the orbicularis oris attaches to the columella, septum, and anterior nasal spine. The lateral orbicularis oris attaches to the alar base and piriform rim. If not completely detached and reconstructed, contraction of the side-to-side repaired muscle may present as bulges on either side of the lip closure.

through proper mobilization and accurate repair of the orbicularis oris muscle. The orbicularis oris is a fanlike structure with a superficial component and a deep component. The former, in association with extrinsic facial muscles, serves as a retractor of the upper lip. The deep component of the orbicularis oris, subdivided into pars marginalis and pars peripheralis, is responsible for the sphincteric action of the mouth. The pars marginalis forms the innermost layer of the oral fissure and is surrounded by the pars peripheralis in a concentric manner.

Abnormalities in muscular anatomy vary, depending on the severity of the cleft lip. The deficiency of the orbicularis is more marked on the medial side than on the lateral side. In incomplete clefts, even though some muscular fibers remain in the upper portion of the lip, these seem to be attenuated and lacking sphincteric action. In the complete cleft lip, the general direction of muscle fibers parallels the cleft margins. The pars marginalis is interrupted on either side of the cleft at the point where the vermilion–cutaneous junction becomes thin. The peripheralis on the lateral side has abnormal insertions to the alar base and the periosteum of the piriform aperture. Similarly, on the medial side, it has insertions to the columella, nasal septum, and anterior nasal spine (► Fig. 13.3).

Mulliken noted that in the normal Cupid's bow, the anterior projection of the pars marginalis gives rise to the white roll or vermilion–cutaneous junction. However, in the affected lip, the pars marginalis was found to be both hypoplastic and disoriented adjacent to the cleft. Furthermore, the vermilion width, though normal on the lateral side, was significantly deficient on the medial side of the cleft lip.

Emphasizing on the importance of the natural-appearing philtral column during cleft lip correction, Mohler (1987) created a classification of philtral configuration. Type 1 is most

common, with some degree of divergence of the philtral column at the base of the columella. Type 2 has a degree of convergence at the base of the columella. Lastly, type 3 has a convergence below the base of the columella.

13.4 Anthropometry of the Lip

Before any attempt to correct a deformity, the norm has to be correctly understood. Farkas has documented the normal anthropometry of the face in detail. Such measurements offer valuable insight into the structural growth of the nasolabial complex. By using this guide, a lip may be constructed to achieve optimal results. It is crucial to maintain photographic and numeric records, so that an accurate assessment can be done on the results of a surgeon's protocol. Newer three-dimensional anthropometry systems are particularly convenient, as they can record accurate measurements in a split second.

Farkas et al (1992) found that cutaneous upper lip height reached approximately 80% of its growth potential by 1 year of age and 94% by 5 years of age. Likewise, upper lip height and nasal and prolabial width show rapid growth early in life. This is in contrast to the columellar length and nasal tip projection, which show slow growth. Measurements should be made to intentionally overcorrect the slow-growing structures, as opposed to the fast-growing structures. Such distinction between fast- and slow-growing nasolabial elements is the key to planning a correction in the fourth dimension. Using anthropometric measurements, Thomas et al (2013) recently reported on normal nasolabial anatomy in infants younger than 1 year (► Fig. 13.4).

13.5 Anatomy of the Cleft Nasal Deformity

Pathogenesis of the cleft nasal deformity can be attributed to three major factors: (1) muscle imbalance, (2) septal deformity, and (3) asymmetry of the skeletal base. The extrinsic muscles of facial expression, attached to the orbicularis oris muscle on the cleft side, are responsible for the lateral pull of the alar base. Contraction of the unopposed orbicularis muscle insertion on the medial side results in pulling of both the septum and the columella to the noncleft side. Further contributing to the septal deformity is the septospinal ligament, which is also unopposed on the normal side, fanning from the caudal border of the septum to the anterior nasal spine. This causes the caudal septum to be hypertrophied and dislocated off the spine, away from the cleft.

Hypoplasia (or posterior location) of the lesser maxillary segment and an asymmetric skeletal base contribute to a generalized displacement of the nasal pyramid toward the cleft side and to the resultant stretching and flattening of the dome of the alar cartilage. The lower lateral cartilage is inferiorly distracted, and its normal overlapped relationship with the upper lateral cartilage is distorted. As a result, a buckling deformity of this cartilage is usually noted. The orientation of the medial and lateral crura of the alar cartilage on the cleft side is also affected. The medial crus is inferiorly displaced on the cleft side,

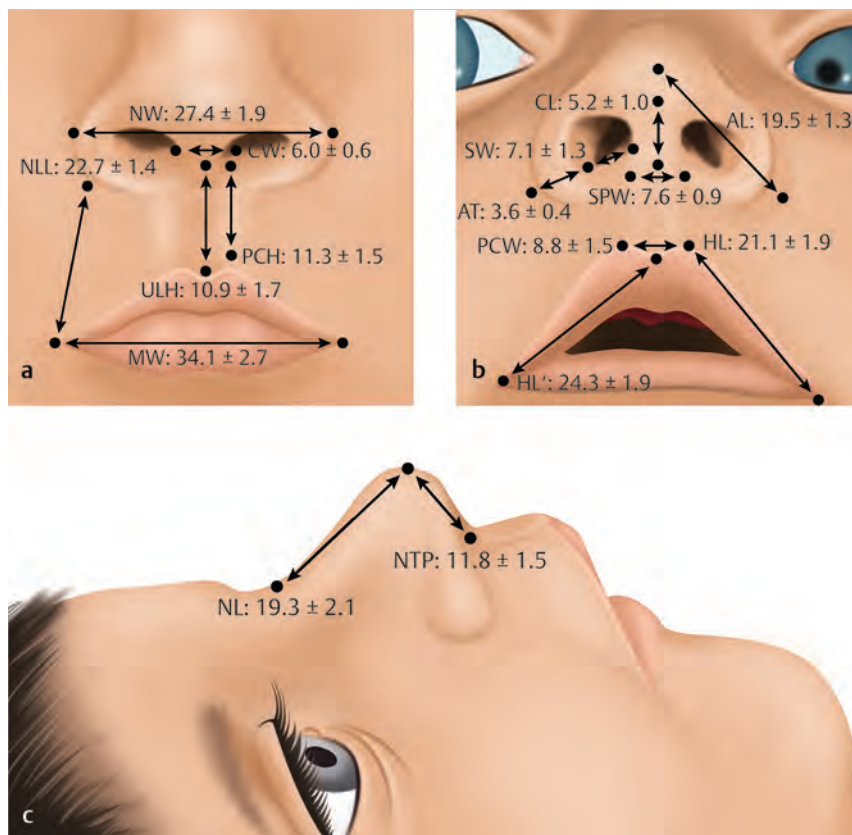


Fig. 13.4 Representation of normal nasolabial anthropometry in an infant.

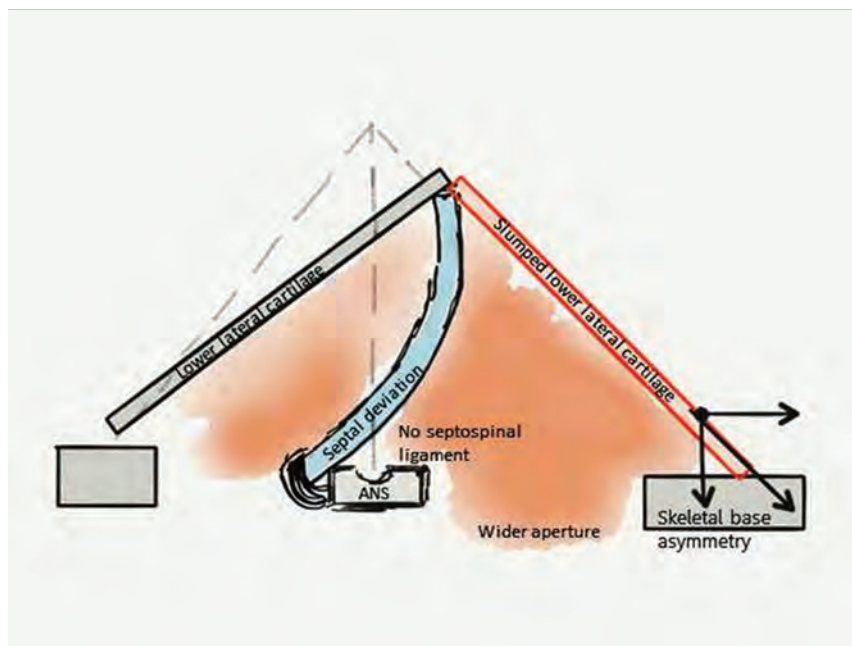


Fig. 13.5 The tripod theory suggests that the cleft nasal deformity is a result of skeletal imbalance, septal dislocation, and width of the cleft and does not self-resolve if uncorrected.

whereas the lateral crus is usually rotated inferiorly (► Fig. 13.5). The nasal vestibular mucosa is also stretched secondary to the width of the cleft. A combination of excess mucosa and displacement of the lateral crus of the lower lateral cartilage can result in a vestibular web after correction of the cleft lip.

13.6 Preoperative Management

13.6.1 Dentofacial Orthopedics

There are a variety of options for presurgical treatment of the alveolus. They vary depending on expertise, accessibility, finances, and individual protocols. There are three basic methods of

presurgical molding: taping, passive molding, and active orthopedic devices. Taping is the simplest method, with external force placed by gradual approximation of the cleft edges of the lip with adhesive tape. A dental plate may be used in conjunction with taping to prevent collapse of the lesser segment. However, accurate alignment is challenging, and dermatitis and epidermolysis from frequent tape removal can be annoying. Alveolar approximation is also possible by progressive molding of a dental plate. The dental plate is held in place with an adhesive and gradually shaved on the palatal surface. External force by taping assists in moving the arches along this molded surface. Since infant cartilage can be molded nonsurgically, a nasal phalange is added when the interalveolar distance is less than 6 mm. This nasal molding bulb is progressively straightened to achieve an excellent preoperative position of the affected lower lateral cartilage. Usually, this process takes 10 to 12 weeks. A simultaneous correction of the lip, nose, and alveolus (gingivoperiosteoplasty) is possible after nasoalveolar molding (NAM).

An active dentomaxillary or Latham appliance accomplishes alveolar alignment much more quickly (4–6 weeks). A custom-made prosthesis is secured to the palate with pins (under a brief anesthetic). A screw is turned weekly to activate an arm that moves the lesser segment anteriorly along the radius of the arm. An auxiliary elastic chain can be engaged to assist in decreasing the interalveolar gap. A stronger force is applied on the maxillary segments for proper positioning. Presurgical orthopedics effectively narrows the cleft, facilitating primary repair and decreasing the need for secondary repair. No long-term effects have been noted with use of dentofacial orthopedics (► Fig. 13.6). Proponents of active dentofacial orthopedics tout the ability to move the lesser segment anteriorly in infants with significant anteroposterior (AP) discrepancy (► Fig. 13.7).

13.7 Operative Treatment

13.7.1 Lip Adhesion

A lip adhesion can be a useful adjunct in the treatment of a complete cleft lip. An adhesion may be used instead of or in conjunction with dentofacial orthopedics. If access to presurgical orthopedics is unavailable, a lip adhesion can be performed as a staged procedure to narrow the cleft and approximate the dental arches, allowing for staged correction of the nasal deformity (► Fig. 13.8). This may decrease the number of secondary procedures necessary. However, many centers have abandoned a preliminary adhesion, given that it may lead to exposure to anesthesia complications and scarring, and may add to cost.

Markings for a lip adhesion are made beyond any landmarks that are important for the eventual cleft lip correction. On the medial side, a linear incision is marked 2 mm beyond the vermilion–cutaneous junction that extends along the base of the caudal septum. On the lateral side, a similar, corresponding incision is marked in the lip (beyond the vermilion–cutaneous junction) to the level of the piriform. A perpendicular incision is marked from the vestibular–piriform margin along the lateral buccal sulcus. The lip incisions are made to the level of the submucosa, until the underlying muscle is visualized. However, the muscle is not dissected. The cheek is elevated along the supra-periosteal plane, so it may be advanced across the cleft for a tension-free closure. A back cut in the mucosa greatly assists in

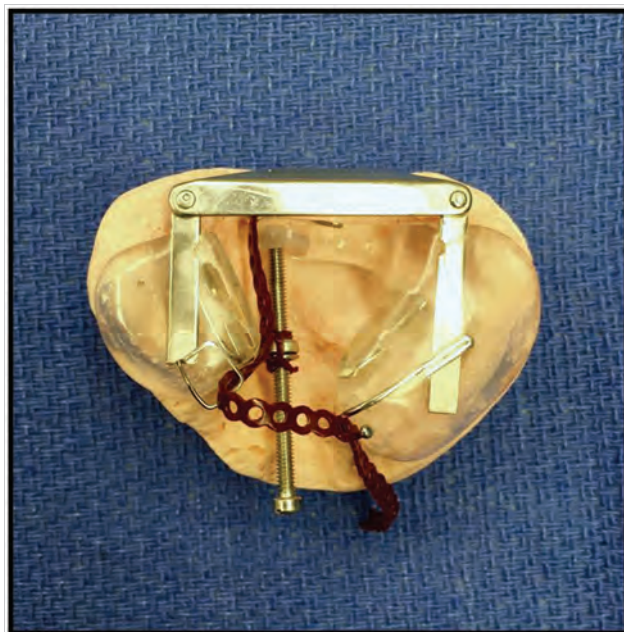


Fig. 13.6 One example of preoperative dentofacial orthopedics is a Latham appliance. It is a pin-retained palatal appliance, which is used to correct the AP discrepancy with the turning of the screw; the width of the cleft is narrowed with tightening of an auxiliary elastic chain.

advancing the flap along the buccal sulcus, and the posterior mucosa of the adhesion is closed. The nasal floor is constructed by apposing the septal mucoperichondrium and the lateral vestibular mucosa. Interrupted sutures are placed in the muscular layer, and the anterior mucosa is carefully approximated. Thin adhesive skin-closure strips (Steri-Strips) reduce the tension across the adhesion in the immediate postoperative period. Typically, the final correction of the cleft lip is delayed for 2 to 3 months after the adhesion (► Fig. 13.9).

13.7.2 Gingivoperiosteoplasty

Efforts to close the cleft alveolus primarily (with bone grafts) may produce detrimental effects on maxillary growth, especially if dissection is aggressive. Primary bone grafting has been abandoned at most institutions. However, an unrepaired alveolar cleft can result in fluid and air leakage. One option to close the cleft alveolus is a gingivoplasty or gingivoperiosteoplasty. After approximation of the arches with dentofacial orthopedics, local gingival tissue can be used with limited undermining to close the alveolar cleft (► Fig. 13.9d). A gingivoperiosteoplasty effectively closes the alveolar cleft and completes the dental arch to provide a stable platform for nasal correction and tooth eruption.

More commonly, the alveolar cleft is left untreated until secondary bone grafting. Typically, secondary alveolar bone grafting is performed at a stage of mixed dentition, when the canine root has not fully formed (age 8–10 years). A bone graft is performed to provide bony support for the teeth adjacent to the cleft and a dental implant (for the missing lateral incisor) or to allow for movement of the permanent canine into the lateral incisor position. Other advantages of secondary bone grafting include stabilization of the arches, provision of gingival contour,



Fig. 13.7 Case example: In cases of significant AP discrepancy in the alveolar arches, the use of dentofacial orthopedics is invaluable. An active appliance promotes anterior movement of the lesser arch. After aligning the arches, a one-stage lip and nasal correction was performed. At 3 years of age, the boy has a slight recurrence of the vestibular web, which may be corrected electively.

and support of the alar base. Secondary grafting is successful by using cancellous bone from the iliac crest.

13.7.3 Controversy of Adjunctive Treatment in Cleft Lip and Nasal Deformity

Anecdotally, the incomplete cleft is the easiest to correct. The dental arches are well aligned, the alar base is in a good symmetric position, and there is greater orbicularis bulk secondary to tension from partial completion of the oral sphincter. In fact, just the presence of a skin bridge across the cleft (Simonsart's band) greatly minimizes the severity of the cleft. Based on these observations, Millard introduced a protocol to convert a complete cleft into an incomplete cleft. A dentofacial appliance is first used to approximate the dental arches. Then, lip

adhesion and gingivoperiosteoplasty are performed to set up the foundation for correction of a complete cleft. This protocol, also popularized by Mulliken, has the advantages of arch alignment, preferred alar base position, and increased muscle bulk, as well as a slight lengthening of the medial element of the lip. This can reduce the length of the back cut necessary for the rotational flap, so that it does not encumber the lip or philtrum on the normal side. Staged correction of the nasal deformity also yields better and more consistent results (► Fig. 13.10 and ► Fig. 13.11).

Alignment of the arches may be performed by either active or passive dentofacial orthopedics. Controversy surrounds the use of dentofacial appliance and the effect of gingivoperiosteoplasty. Supporters refer to the advantages of not having an alveolar fistula and obviation of a secondary bone graft, without significant effect on maxillary growth. Detractors point to its

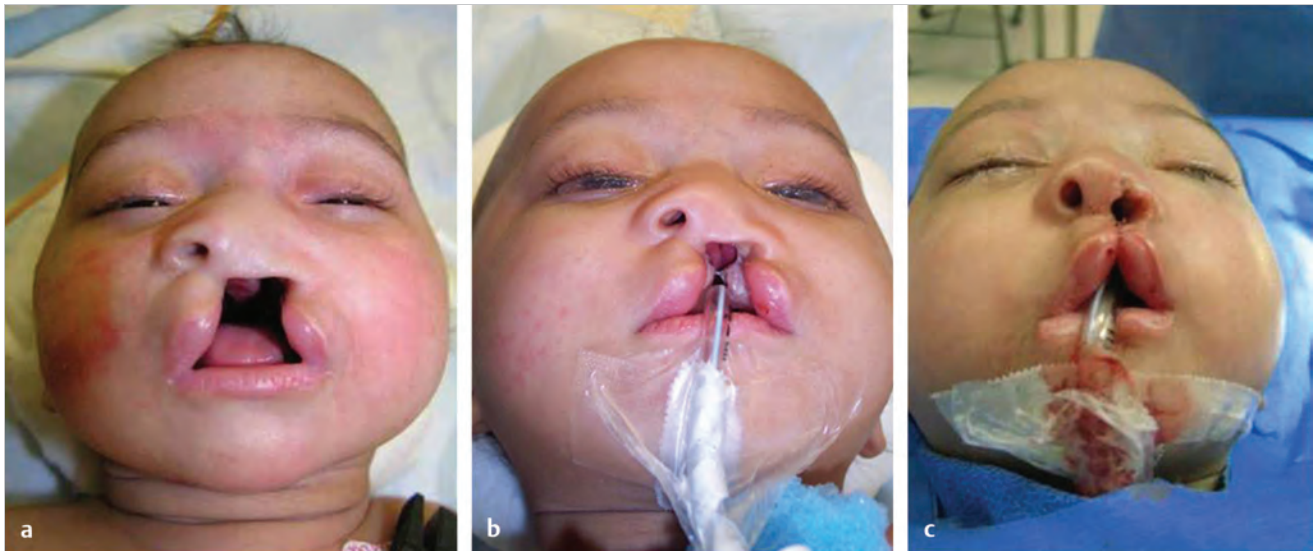


Fig. 13.8 Converting a complete cleft into an incomplete cleft, using dentofacial orthopedics and lip adhesion, results in less tension across the eventual correction, increased medial lip height, a better foundation for lip and nasal correction, improved alar base symmetry, and more muscle bulk at the site of repair. It also allows for staged correction of the nasal deformity.

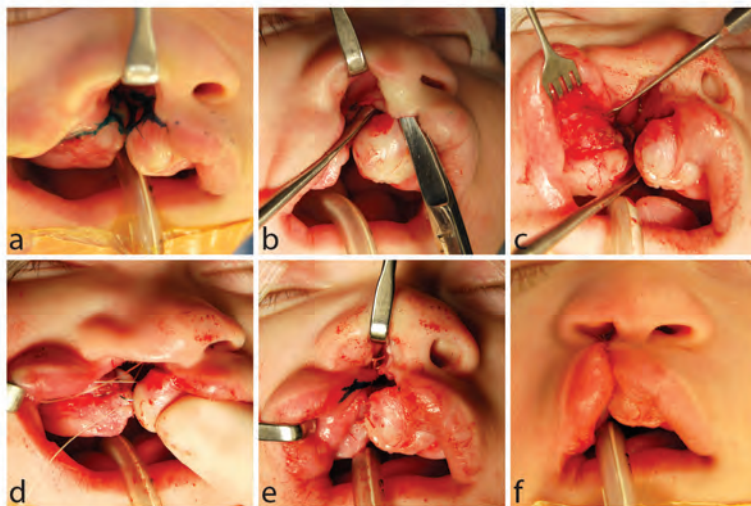


Fig. 13.9 (a) Markings for a lip adhesion. (b) Medial dissection involves raising a septal mucoperichondrial flap, as well as separating the abnormal intercrural attachments in the columella. A limited dissection of the lip, only to visualize the muscle, is performed. (c) Lateral dissection involves a buccal sulcus incision, supra-periosteal dissection over the maxilla, as well as raising the lateral vestibular mucosa off the piriform rim. (d) Closure begins with a back cut in the buccal sulcus incision and closure of the sulcus. Finger pressure, along with a medial incision in the alveolus, can assist in closure of the gingival gap. (e) The nasal floor is constructed by approximating the septal mucosa to the lateral vestibular mucosa. The buccal mucosa of the upper lip is also approximated. (f) Finally, the orbicularis is apposed in a side-to-side fashion with three interrupted sutures, and the mucosa lining the cleft is closed. All key landmarks required for the eventual lip and nasal correction are untouched during this operation.

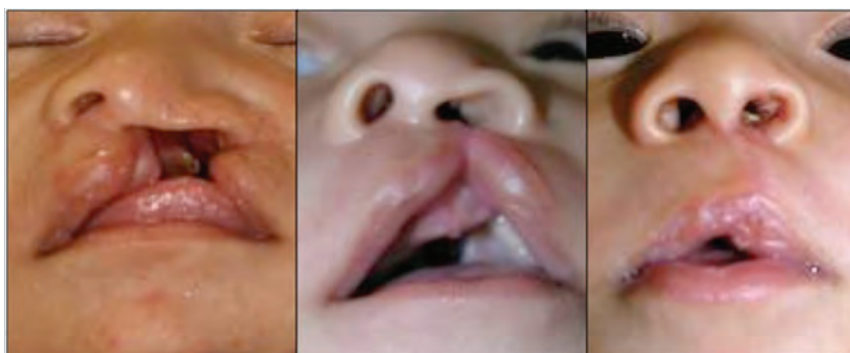


Fig. 13.10 Staged correction of the cleft nasal deformity yields more consistent symmetry.



Fig. 13.11 Case example: Example of a boy with a left unilateral complete cleft lip and palate, who underwent a two-stage lip and nasal correction, followed by a palate closure. At 6 years of age, he is starting school, with no other revisions necessary.

effect on restricting maxillary growth and an abnormal bite relationship. This is most likely secondary to the closure of the lateral incisor space and restriction of the sagittal growth of the maxilla. However, recent studies have documented that a cleft of the primary palate, rather than operative strategy, is the inciting cause of maxillary restriction. In the same vein, inherent differences in health care systems, treatment protocols, and ethnic expectations, as well as cultural acceptance of concavity of the facial profile, have led to inadequate elucidation of treatment outcomes.

13.7.4 Surgical Repair

Markings

Important landmarks of the lip and nose should be marked, such as the subnasale (sn), columella (c), subalar point (sbal),

and Cupid's bow peak (cphi). On the lateral side, sbal is marked on the affected side (► Fig. 13.12). Note that the subalar point will be splayed laterally secondary to the under-rotated alar base of the cleft nose (► Fig. 13.13d). In order to obtain a desirable result, an equal vertical length of the cutaneous upper lip must be achieved. The most important point to mark is the Cupid's bow peak (cphi) on the lateral side. Landmarks unaffected by the cleft are the sbal and the cphi on the unaffected side. Therefore, this length should serve as a template for the affected side. Calipers are useful to first determine the length of the upper lip on the unaffected side (sbal to cphi) and then to mark the cphi on the affected side by marking the point where the arc of the length of the lip meets the vermilion–cutaneous (white roll) junction (► Fig. 13.14). The horizontal length between the cphi and its ipsilateral commissure or chelion is recorded. However, it has been shown that a short preoperative

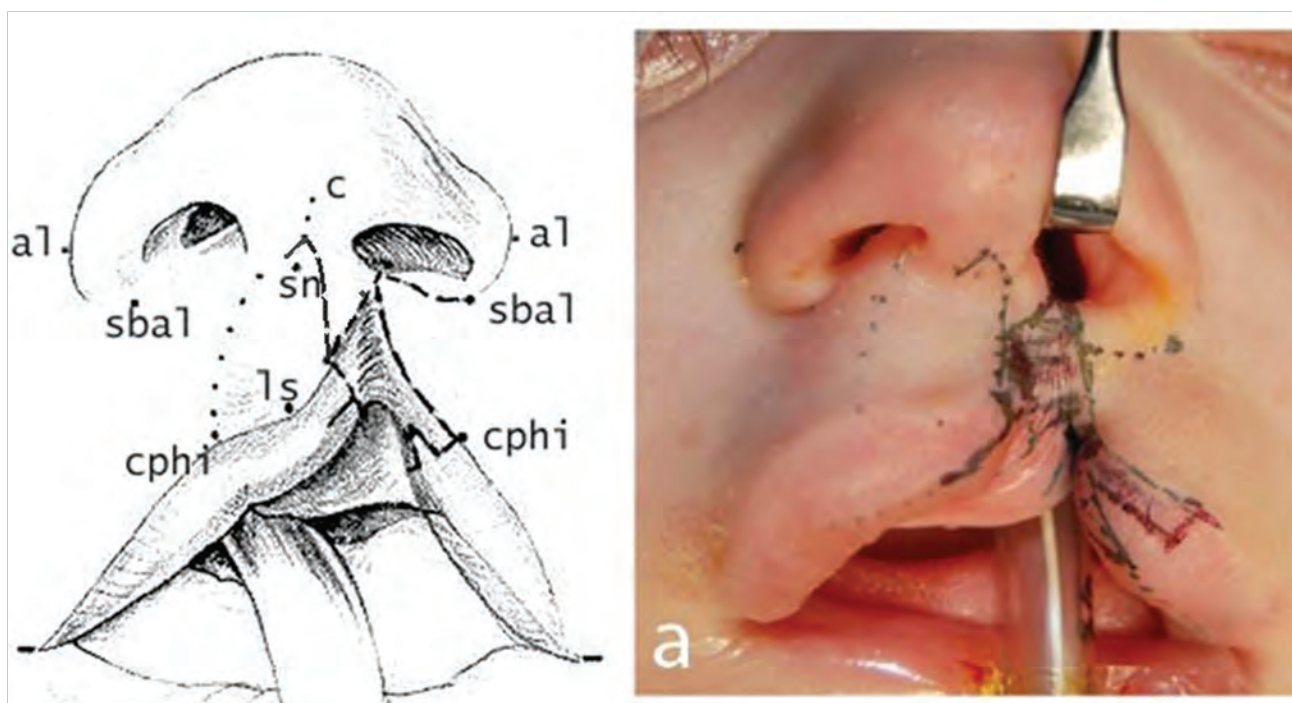


Fig. 13.12 Markings for a cleft lip correction. Key landmarks are first noted. Based on this, a rotation flap that mirrors the opposite philtrum is marked. It extends into the columella, where a Mohler-type back cut is marked. This allows for the hemicolumella to be retrogressed to a more anatomic position. The incision is prevented from entering into the lip, which would be visible as well as cause an effacement in the superior aspect of the philtrum. The advancement flap is marked from the proposed Cupid's bow peak into the nasal floor. Although the perialar incision is unnecessary in incomplete clefts, it is marked in case greater cutaneous length of the upper lip is necessary for symmetry. In cases of a long upper lip, a perialar incision would obviate the need for the Cupid's bow peak to be very lateral on the upper lip, which may create asymmetry from the commissure to the philtrum.

horizontal distance lengthens after correction of the complete oral sphincter. The distance between cphi and labiale superius (ls) is used as a guide to mark the proposed cphi on the medial side.

Markings can then be made for the rotation and advancement flaps (► Fig. 13.12 and ► Fig. 13.13c). The rotation flap is marked along the medial margin of the cleft, mimicking the unaffected philtrum and curving toward the columella. A back cut is designed at the top of the rotation flap (► Fig. 13.13d). Based on the convergence or divergence of the opposite philtrum, the back cut may be trimmed for symmetry during closure. The advancement flap is marked on the lateral lip element from the proposed cphi to the nasal sill along the cleft margin and may be extended to the alar base, as in the case of a complete cleft (► Fig. 13.15). The columellar flap (C-flap) is marked from the superior edge of the rotation flap to the membranous septum. This flap will be used to augment the hemicolumella and reconstruct the nasal sill (► Fig. 13.13e).

Attention then turns to the vermillion and mucosa of the lip. The vermillion is deficient on the medial edge of the cleft; therefore, a triangular flap of vermillion is designed at the vermillion-mucosal junction ("red line"). The labiobuccal mucosa is also deficient; a triangular flap of mucosa is planned from the lateral lip to elongate it. A gingivobuccal incision is planned from the cleft edge to raise the cheek flap for advancement across the cleft (► Fig. 13.13g). A third triangular flap carrying the white roll will be designed just before closure of the philtral incision. The late use of the triangular flap allows for correction of

submillimeter differences that may be present after the rotation-advancement repair has been completed. It also breaks up a linear scar vertically across the lip and prevents scar contracture, especially in the vermillion-cutaneous junction (► Fig. 13.16).

Dissection

Incisions are made along the markings, and the mucosa lining the cleft is discarded. The buccal sulcus incision is made; however, it does not extend to the level of the parotid duct. A supra-periosteal dissection is performed on the maxilla up to the level of the infraorbital foramen. In case of an incomplete cleft lip or if a lip adhesion has been performed previously, extensive maxillary dissection is not necessary. Attention is then turned to the lip. The orbicularis is first separated from its anomalous attachment to the alar base and then dissected away from the skin and mucosa of the lip (► Fig. 13.13h). A mucosal back cut in the buccal sulcus assists in advancing the lateral lip elements across the cleft (► Fig. 13.13g). Scissors are placed between the medial crura (under the columellar flap), and blunt dissection is used to separate it from the opposite medial crura and dome. A sharp back cut at the top of the rotation flap (on the columella, toward the opposite philtral column) is then created while the affected nostril is held at an optimal level with a retractor. The back cut is performed until the Cupid's bow peaks are level (► Fig. 13.13e). If not addressed during a lip adhesion, the septal mucoperichondrium and the lateral vestibular mucosa are elevated for nasal floor reconstruction.

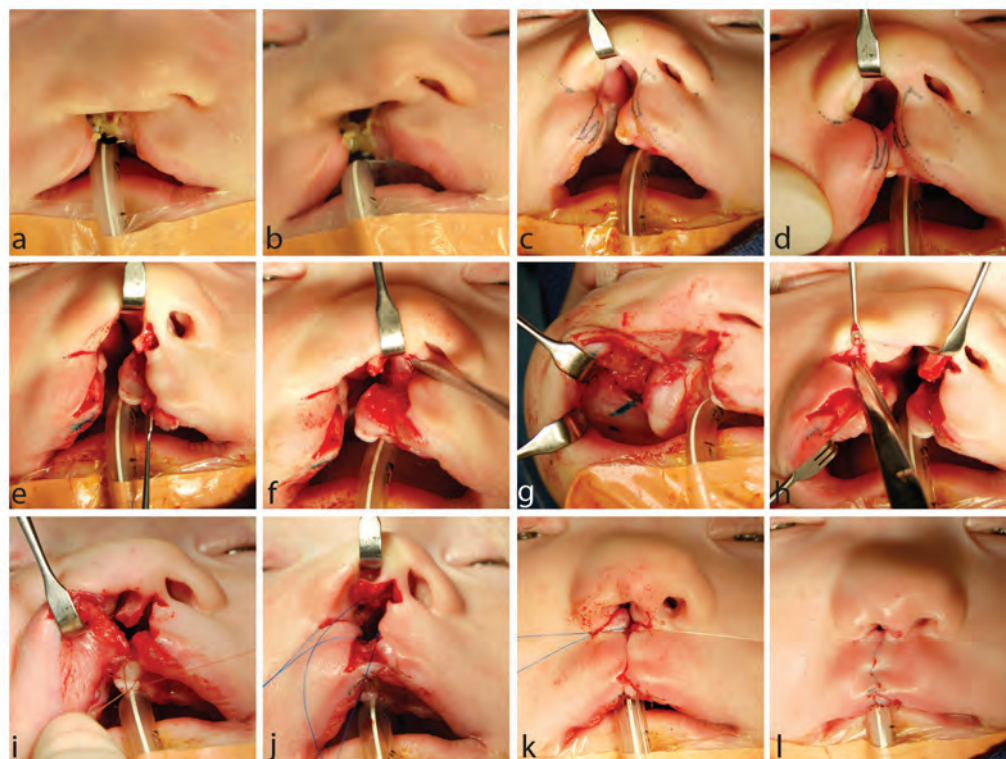


Fig. 13.13 (a,b) A right unilateral complete cleft lip and nasal deformity after alveolar alignment with active dentofacial orthopedics. (c) Markings for a rotation-advancement repair. (d) Medialization of the lateral element will assist in accurately siting the subalar point (sbal). (e) The mucosal lining of the cleft is excised. The back cut above the rotation flap is restricted to the columella. (f) Note the dislocation of septum to the noncleft side of the spine. It is relocated and secured to the cleft side of the anterior nasal spine. The septal mucoperichondrium will be used to construct the nasal floor. (g) A mucosal back cut in the buccal sulcus assists in advancing the cheek and lip across the cleft, without any tension on the eventual closure. The maxillary dissection is in the supraperiosteal plane. The lateral vestibular mucosa is raised off the piriform rim to construct the nasal floor. (h) The orbicularis muscle has been separated from its cutaneous and mucosal attachments. Abnormal attachments of the muscle to alar base and columella are separated. (i) The buccal sulcus incision is advanced medially and closed, as the leading edge is sutured to a releasing incision near the frenum of the medial upper lip. A linear incision along the cleft margin of the alveolar gingiva is made, and the lingual and buccal portions of the alveolus are approximated. This is possible only if the alveolar segments are adjacent to one another from presurgical orthopedics. (j) A guy suture from the maxillary periosteum is placed into the alar base, to be tightened later. The medial crura of the lower lateral cartilages are approximated under the C-flap. This will allow retrogression of the C-flap into the back cut in the columella. The laterally based vermilion flap is inserted into a releasing incision along the vermilion-mucosal junction medially (which is deficient in all cases). (k) The orbicularis oris muscle is apposed from an inferior to superior fashion, with increasing tension superiorly. The superior-most portion of the orbicularis is tacked down to the anterior nasal spine in order to recreate the pout. The alar base is sutured to the maxillary periosteum, as well as to an appropriate level of the underlying orbicularis muscle, so as to accurately secure the alar base in a symmetric position. (l) The skin of the upper lip is draped over the muscle closure. The rotation flap is trimmed to obtain a symmetric philtral column. Along the inferior border of the rotation flap, a releasing incision lowers the Cupid's bow to a symmetric position. A small triangular flap from the advancement side, carrying the white roll, is inserted into this defect, and the rest of the philtral incision is closed. The superior edge of the advancement flap is tucked into the nostril to recreate the morphology of a normal nasal sill.

Closure

The nasal floor is closed by approximating the septal mucoperichondrium and the lateral vestibular mucosa. While retracting the nostril to a level symmetric to the contralateral side, the columellar flap is retrogressed into the hemicolumella. The alar base flap is rotated endonasally and is approximated (end to side) to the columellar flap to form the nasal sill. The buccal sulcus is closed after advancing the back cut. A triangular mucosal flap at the leading edge of this flap is interdigitated into the labial frenum to augment the mucosa on the cleft side (► Fig. 13.10i). The buccal side of the lip mucosa is then closed. The orbicularis oris is then apposed from an inferior to a superior accession. The most superior portion of the orbicularis is

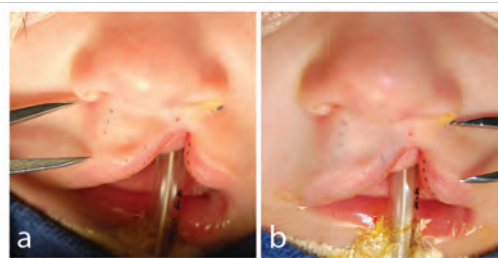


Fig. 13.14 Sighting the proposed point for the Cupid's bow peak. Calipers are used to measure the length of the cutaneous upper lip on the unaffected side (sbal-cphi), and this distance is used to mark the proposed peak along the vermilion-cutaneous junction.



Fig. 13.15 Case example: In cases of a long upper lip on the unaffected side, it may be necessary to perform a perialar incision in order to obtain a longer cutaneous length for the affected side. This prevents the need to mark the Cupid's bow peak in a more lateral position that may result in greater asymmetry. It also allows for independent positioning of the alar base. Satisfactory lip and nasal correction is seen at 5 years of age. The perialar incision has healed well.

affixed to the anterior nasal spine in order to re-create the appearance of a pout (► Fig. 13.13k).

The alar base is secured to the underlying periosteum of the medial maxilla deeply and to orbicularis muscle inferiorly, to a point where the interalar distance is symmetric in all axes (► Fig. 13.13j). A 1-mm overcorrection is prescribed, as post-operative widening is generally noted. The cutaneous closure of the lip starts at the vermilion–cutaneous junction. A small triangular flap (the white-roll flap) is inserted into a releasing incision on the medial side to complete the white roll and dictate the balance of the Cupid's bow (► Fig. 13.13l). The triangular flap of vermilion is inserted into a releasing incision along the vermilion–mucosal line (wet-dry junction or red line) to complete closure of the mucosa of the upper lip. The advancement flap is then trimmed to a convex shape and is closed to the rotation flap in an inferior-to-superior direction (► Fig. 13.16). The subnasal portion of the rotation flap is trimmed according to the shape of the philtrum and closed along the nasal sill. The distal edge of the advancement flap is trimmed and inserted into the superior portion of the philtral incision, without any tension.

Cleft Nasal Correction

In spite of lip correction, the nose usually demonstrates a “recurvatum” deformity with buckling of the lower lateral cartilage (► Fig. 13.17a). Primary correction of the nasal deformity is prescribed in all clefts. Internal resorbable sutures are recommended. The use of percutaneous sutures yields a nonanatomic correction, and the use of bolsters is inadequate, since they are usually removed in a week (cartilage healing takes considerably longer).

For adequate correction, an infracartilaginous incision is performed, exposing the lower lateral cartilage (► Fig. 13.17b). Using a cotton-tipped applicator, the contralateral dome is delivered through the marginal incision; an interdomal suture is placed between this spot and the proposed dome of the affected cartilage. The lateral crus of the lower lateral cartilage is suspended from the ipsilateral upper lateral cartilage to reconstruct the scroll (which is usually separated due to the stretch across a complete cleft; however, it may be intact in an incomplete cleft and therefore unnecessary) (► Fig. 13.17c). Laterally, soft tissue attachments to the piriform displace the

lateral crus of the lower lateral cartilage. Division of these attachments allows the cartilage to assume a normal position. Correction of the lower lateral cartilage back to the normal position usually results in redundancy or “webbing” of the vestibular mucosa and warrants a lenticular excision and suspension to open up the nostril. Overcorrection is recommended to overcome the perverse tendency of the lower lateral cartilage to relapse (► Fig. 13.17d).

Owing to incisions along the columella, nasal floor, alar base, and nostril roof, a circumferential contraction of scar can lead to nostril stenosis. To avoid nostril stenosis, a postsurgical

silicone conformer can be used during the healing phase. However, an external splint may not be tolerated by infants and may also cause frictional abrasions and pressure-related ulcerations.

13.8 Microform Cleft Lip Correction

A microform cleft lip, as described by Onizuka et al, includes (1) minimal nasal deformity, (2) philtral groove, (3) thin free margin of the lip, (4) notch at the vermilion–cutaneous junction (white roll), and rarely (5) notch in the alveolus. It is important to note that the philtral groove is an indication of an incomplete oral sphincter and should be corrected during the operation. Correction of the microform cleft lip can be done either by a rotation–advancement repair (for severe defects) or by a “double unilimb Z-plasty.” Triangular flaps at the vermilion–cutaneous junction and the vermilion–mucosal junction are used to augment the medial element by inseting the flaps into corresponding releasing incisions. The orbicularis is carefully apposed, and the alar base is endonasally rotated in a Y–V advancement. Lastly, correction of the slumped lower lateral cartilage is performed as previously described (► Fig. 13.18).

13.9 Postoperative Care

There is no evidence to support the use of arm restraints during the postoperative period in children undergoing cleft lip or palate surgery. Although the use of a syringe for feeding has been proposed, in our practice, both bottle- and breastfeeding are allowed. Postoperative taping is done for 3 to 6 weeks, and gentle scar massage is recommended thereafter. Scar creams can be prescribed, but there is no conclusive evidence for any aesthetic benefit.

13.10 Technical Differences in Unilateral Cleft Lip Correction

After its original description by Dr. Ralph Millard in 1955, the rotation–advancement technique has remained a basic foundation for unilateral cleft lip correction in cleft centers throughout the world. Several modifications have been described based on

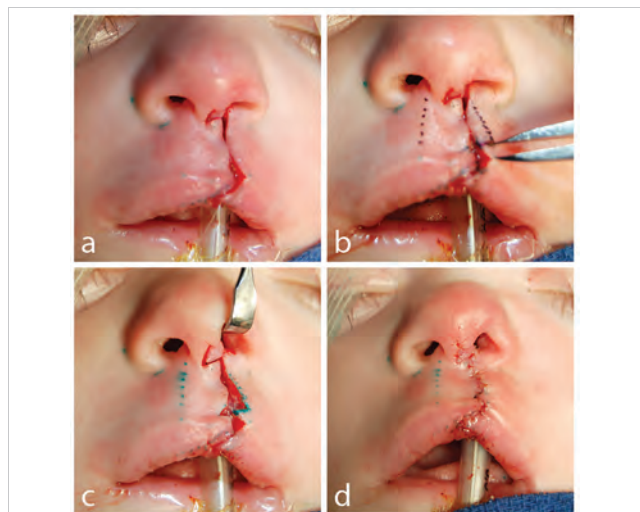


Fig. 13.16 Demonstration of a Tennison's triangular flap at the Cupid's bow. (a) When the back cut is limited from encumbering into the lip, there is usually a deficiency in the Cupid's bow peak at rest. (b) The difference is measured and an equilateral triangle is marked in the inferior edge of the advancement flap. The rest of the flap is trimmed (usually the least vascular portion of the flap). (c) A releasing incision at the vermilion–cutaneous junction on the medial side lowers the Cupid's bow peak to a symmetric level. (d) The triangular flap (carrying the white roll) is then inset, and the rest of the philtral incision is closed. Note that a second triangular flap is used to augment the vermilion deficiency that is usually present on the medial element of the cleft lip. These triangular flaps break up a linear vertical scar and prevent contracture, especially at the level of the vermilion–cutaneous junction.

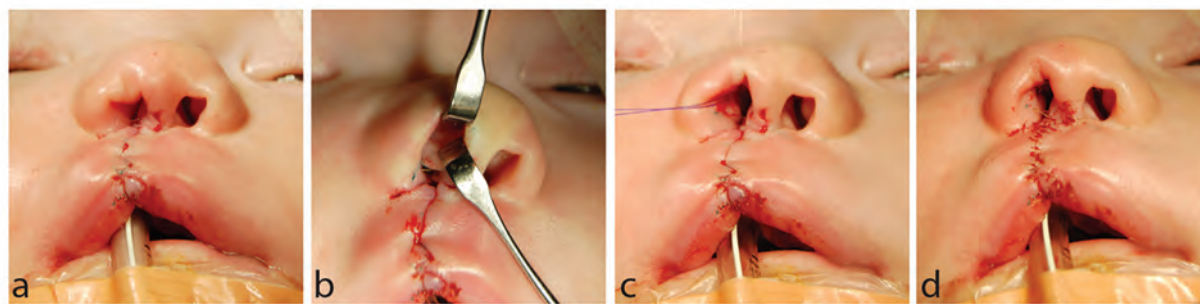


Fig. 13.17 Primary correction of the cleft nasal deformity. (a) A “recurvatum” deformity of the lower lateral cartilage is noted, in spite of the correction of the alar base, septum, and medial crura (via the back cut). (b) Exposure of the upper and lower lateral cartilages via a closed approach. The lateral soft tissue attachments of the lower lateral cartilage are released. (c) Interdomal and lateral crural suspension sutures are placed. (d) Slight overcorrection is prescribed due to the perverse tendency for the deformity to recur.

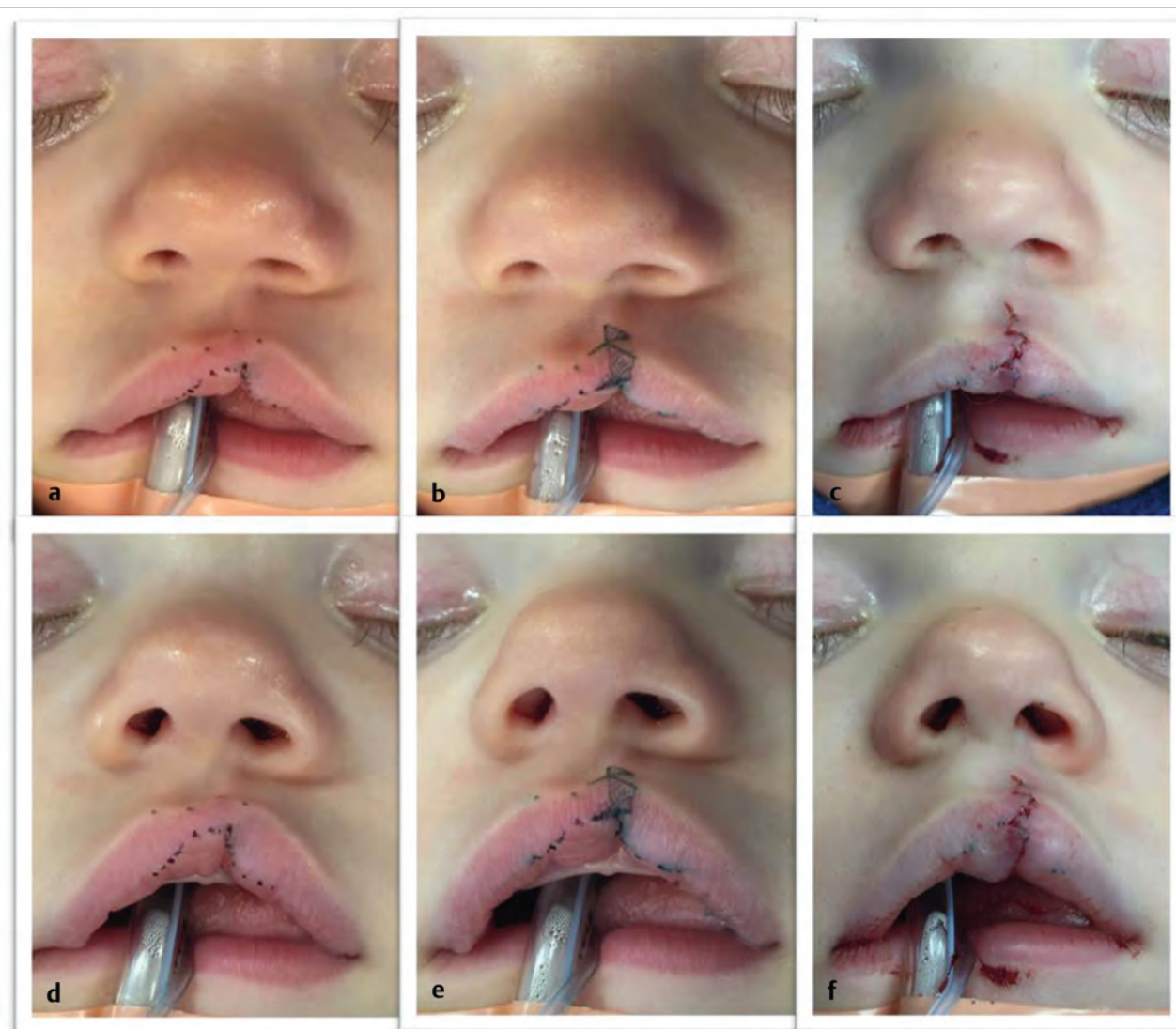


Fig. 13.18 Correction of a microform cleft lip. (a,b) AP and submental views of a microform cleft lip and nasal deformity. (c) Final closure. (d) Markings for correction. The vertical deficiency of the Cupid's bow peak is noted, and an equilateral triangle of those dimensions is marked above the proposed Cupid's bow peak. A second such triangle may be required if a vermillion deficiency is present (double unilimb Z-plasty). The intervening tissue is marked for excision. The alar base is marked for a Y-V type endonasal advancement. (e) Dissection in the subcutaneous tissue extends up to the nasal floor and into the submucosa of the upper lip. The intervening fibrous band in the muscle is excised, and the orbicularis is apposed. Endonasal rotation of the alar base and correction of the lower lateral cartilage with a single interdomal suture are performed. A dermal matrix or dermal graft may be used to augment the mucosa of the free margin of the lip as well as to "create" a philtral column. (f) Final closure.

the same original principle. Millard himself proposed the first modification by adding a back cut at the end of the rotation incision, allowing further rotation of the medial element and less tension on the repair (► Fig. 13.19b). In 1987, Mohler further modified this technique by reducing bowing of the incision toward the cleft and extending it up on to the columella, resulting in a wider C-flap to fill the entire medial defect (► Fig. 13.19c). Other modifications have been described for gaining length in the medial lip element, with Mulliken bowing the incision toward the cleft in a higher curve and Stal using an S-shaped incision (► Fig. 13.19d). Fisher (2005) avoids the back cut altogether, relying on the releasing incision at the vermillion-cutaneous junction to obtain the desired length on the medial element (► Fig. 13.19e). Incisions are carefully planned

between the columella, philtrum, and Cupid's bow, thereby treating each subunit of the lip separately.

Many surgeons continue to use the back cut as originally described by Millard. Mohler's back cut is restricted to the columella, thus preventing extension into the lip. Mulliken proposes a variation of this technique, using a "releasing incision" that is perpendicular to Millard's back cut, preventing the rotational incision from either crossing the midline or extending into the lip (► Fig. 13.19d). The C-flap can then be used for hemi-columellar lengthening. Furthermore, it prevents the tip of the advancement flap from crossing and effacing the philtral column. The back cut is avoided altogether by some surgeons, championed by Nakajima and, more recently, by Fisher, releasing incisions above the philtrum and at the level of the Cupid's

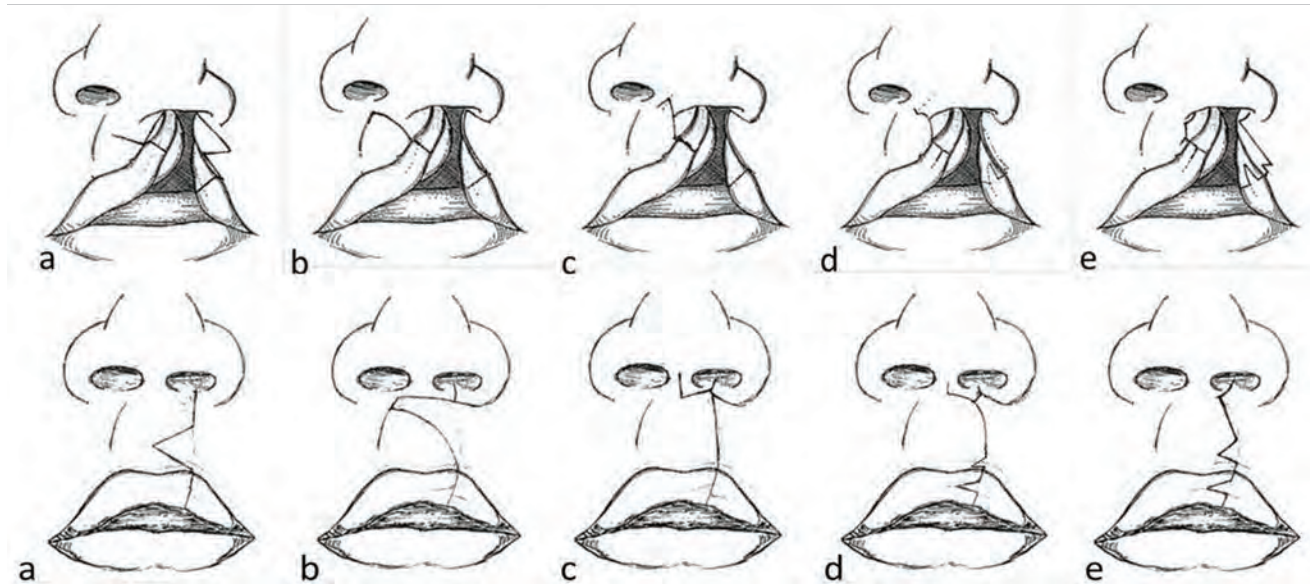


Fig. 13.19 Technical differences in cleft lip repair. (a) Tennison–Randall repair. (b) Millard repair. (c) Mohler repair. (d) Mulliken repair (e) Fisher repair.

bow allows for the correction of the medial segment. Triangular flaps are used to fill these defects from the advancement side.

Extending the incision around the alar base was initially described in the rotation–advancement technique to allow a more independent positioning of the alar base and increased length of the cutaneous upper lip. Detractors of this approach claim that an incision around the alar base could stimulate scar tissue, consequently narrowing the nostril and creating an unattractive horizontal scar. A small perialar–crescentic excision can provide more cutaneous length for the upper lip, without having to lateralize the proposed Cupid’s bow peak. This can be especially helpful in infants with a long upper lip or if the lateral lip discrepancy is great (► Fig. 13.15).

Originally proposed to form the nostril sill and to augment the columella, variations of the C-flap have also been suggested. Mulliken elevates and includes the medial crural footplate with the C-flap by retrogressing it up the hemicolumella. Cutting and Stal advocate the use of a wider C-flap, which when used to backfill the entire medial rotational defect results in a straight-line closure along the nasal sill.

Techniques for muscle repair have significantly evolved from Millard’s original description, where the orbicularis is repaired end to end. Both Cutting and Stal begin by advancing the muscle from the tip of the lateral flap into the medial muscular defect, correcting orbicularis orientation and still allowing a straight-line cutaneous closure. Mulliken begins the repair inferiorly and works superiorly, progressively tightening the closure, resulting in a natural pout of the lower portion of the lip, with the final, superior muscular suture, including the anterior nasal spine periosteum.

13.10.1 Anesthetic Considerations

Several animal studies have shown an association between early exposure to anesthetic agent administration and neurotoxicity. Long-term deficits in learning and cognitive behavior

after early exposure have been noted across several species. Studies on humans have been less consistent. Some studies have suggested that there may be adverse effects on behavior, learning, and memory from prolonged or repeated exposures to anesthesia and surgery when children are younger than 2 to 4 years, presumably before myelination of neural tracts. Some reports have implied a possible association between a single anesthetic exposure and cognitive and behavioral deficits, whereas other reports fail to show any association or show an association only after multiple exposures. Such inconclusive evidence has led to the conception of two large, multicenter prospective human studies, currently under way.

It is still unclear whether it is the duration of a single anesthetic exposure or multiple separate exposures that may lead to worse outcomes. Nonetheless, there is enough evidence in current animal and human studies to be concerned. Although correction of the cleft lip and palatal anomalies is elective, the staging and duration of exposure to anesthesia can be titrated. The cleft or craniofacial surgeon needs to take this into account and be prepared to counsel parents when discussing the operation in an infant with a cleft.

13.11 Complications

13.11.1 Secondary Deformities of the Lip

It is rare that final surgical results do not reflect the careful planning and correct technique used to repair a cleft. However, it is the rule that thoughtless planning and incorrect technique lead to secondary deformity. It is important to evaluate the landmarks of the lip and nose and also to take account of previous procedures, to be able to correct a secondary deformity. Common secondary deformities of the lip can be classified into mucosal, vermilion, Cupid’s bow, orbicularis, cutaneous lip scars, philtral, and nasal deformities.

13.11.2 Mucosa

Mucosal variance (thickness or weakness of the free margin of the upper lip) is very common, given that it is difficult to predict the fullness of the lip during the primary repair. A transverse excision along the vermilion–mucosal junction can address fullness. The scar can be hidden along the red line. Thinness of the free margin of the upper lip can be addressed by augmenting with a dermal fat graft, acellular dermis, or fat injection. Dermal fat grafts are reliable and predictable and are the preferred method; they can be performed in conjunction with other procedures, such as alveolar bone grafts.

13.11.3 Vermilion

Minor step-offs, mismatch, and notching are also encountered. Notching, usually secondary to scar widening, can be corrected by a simple diamond-shaped excision. Step-offs or mismatch at the vermilion–cutaneous junction (white roll) is typically from incorrect alignment during primary repair. A traditional Z-plasty or a unilimb Z-plasty can be useful to correct these deformities. Vermilion deficiency or step-off is encountered if there has been no effort to equalize the width during the primary repair. A triangular flap of vermilion from the wider side can be inset into a releasing incision at the vermilion–mucosal junction to achieve symmetry.

13.11.4 Philtrum

Adult philtral columns narrow under the columella (6 mm) and widen at the Cupid's bow (10 mm). The normal length of the philtrum is 15 to 17 mm. During primary repair (age 3–5 months), the Cupid's bow is 6 mm wide and 6 mm long. The philtrum and the philtral column scars usually widen after repair. Therefore, it is critical to plan the correct shape and size of the philtrum during primary repair. Abnormal shape and size should be corrected to age-appropriate norms.

13.11.5 Cupid's Bow

An upswing of the Cupid's bow, in a corrected unilateral cleft lip, is usually an indication of inadequate rotation of the medial element during primary repair. If the upswing is limited to 1 to 2 mm, it may be corrected with a traditional Z-plasty or a unilimb Z-plasty (Tennison's triangle). If the discordance is worse (short lip), a takedown and a rerotation will be required.

13.11.6 Orbicularis Oris

The continuity of the oral sphincter is paramount in the functional repair of the cleft lip. The orbicularis oris has two parts: the pars peripheralis and the pars marginalis. The pars peripheralis is the functional oral sphincter; the pars marginalis is present in the vermilion–cutaneous junction or Cupid's bow. The most superior portion of the pars peripheralis attaches to the anterior nasal spine. The pathology of the cleft lip results in reorientation of muscle fibers that aberrantly insert into the alar base rather than forming a continuous sphincter. Failure to reorient the muscle by apposing the edges across the cleft may result in a secondary deformity, that is, the contraction of the

muscle during animation, resulting in bulges under the alar base. Failure to tighten and anchor the superior portion of the pars peripheralis to the anterior nasal spine results in a flat upper lip. Inadequate muscle bulk across the cleft can become thin, allowing the philtral scar to contract. Incorrect approximation of the pars marginalis can result in white-roll abnormalities and notching. If the orbicularis discontinuity is suspected, it is necessary to reopen the previous repair and appropriately repair the muscle.

13.11.7 Lip Scars

The appearance of the philtral scar is dependent on the tension across the cleft repair as well as on individual response to injury. It is important to note that philtral scars undergo some contraction immediately after repair. Parents must be counseled to perform scar massage, and, typically, the lip will be balanced after 3 months. Despite best efforts, scars may become hypertrophic, in which case, they may have to be revised. A short lip is usually a result of inadequate rotation of the medial elements during the initial correction. A takedown of the original repair and a rerotation are required. A long lip was traditionally seen after a geometric (triangular or quadrangular) repair of the cleft lip. This is not seen with the advent of the rotation–advancement repair. However, it can be encountered after the correction of an incomplete cleft lip. A complete revision, with suspension of the buccal sulcus and crescentic excision along the alar base, is required to correct a long lip.

13.11.8 Secondary Deformity of the Nose

Unlike secondary lip deformities, which usually result from inappropriate or inadequate correction, secondary nasal deformities are the rule, despite appropriate and adequate correction. The splayed alar cartilage that accompanies a complete cleft lip retains the memory of its fetal position and reverts (to some extent) in spite of the correction. Furthermore, the lack of septal retaining ligaments on the side of the cleft allows the deformity to progress throughout childhood and adolescence. Therefore, it is important not only to correct the nasal deformity primarily but also to revise it in childhood and adolescence to maintain the correction. Similarly, it is important to realize that improper secondary correction can lead to tertiary problems. The dogma of waiting until adolescence to correct the cleft nasal deformity is applicable only for osteotomies.

The tripod theory implies progressive nasal tip and septal deviation secondary to asymmetric displacement of the alar base. Septal deformity and abnormal tension of the cartilage due to malalignment of muscles and the skeletal base (maxilla) can lead to the cleft nasal deformity. It is important to remember that the lower lateral cartilage, although misplaced, is inherently normal anatomically and histologically. The unilateral cleft nasal deformity is secondary to an inferolateral displacement of the medial and middle crura of the lower lateral (alar) cartilage as well as a hemicolumellar deficiency.

McComb and Potter first recommended primary sutural correction of the lower lateral cartilage to prevent progressive deformity. Later studies showed that primary correction of the nasal cartilage did not affect growth. Many adjuncts have been

suggested for maintaining the nasal correction. Cartilage can be shaped permanently if molded in the first few months of life. Preoperative NAM has utilized this principle to obtain superior cleft nasal correction and maintenance. Postoperative nasal conformers can be effective but may be cumbersome and cause columellar excoriation. The use of an internal, resorbable splint to maintain the sutural correction of the lower lateral cartilages has been successful in reducing the need for revisions.

In the quest for ideal position and symmetry, primary correction followed by timely revisions can be more effective than delaying nasal correction until adolescence. Gilles and Kilner stated that a “well-mended harelip would go unnoticed at a cocktail party were it not for the nose.” When we are judging operative correction of a secondary lip or a nasal deformity, it is also important to determine the impact on the patient. In other words, substantiating a qualitative improvement rather than a quantitative improvement should be proven to the child, especially in lieu of medical, academic, social, familial, and financial obstacles. Marsh stated that the surgeon is often more critical of the result than the child with a cleft. Therefore, it is important to know “when enough is enough” in the search for the ideal.

13.12 Conclusion

In parts of Africa, children with clefts are discarded as cursed souls. An operation to correct the appearance of the face can change the lifeline of these children. There are so many important functional considerations such as feeding, hearing, speaking, and chewing that are necessary for children with cleft lip and palate. However, appearance seems paramount, as explained by William Mayo's oft-quoted canticle: “It is the divine right of man to look human.” Repairing the cleft not only addresses the appearance and function of the lip but also heals the psyche of the afflicted. Dr. Joseph Murray, Nobel laureate, perhaps explains it best: “I prefer to think of our work as operating on the soul.”

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14 Bilateral Cleft Lip

Alexander C. Allori

Summary

Repair of bilateral cleft lip has been regarded by many to be among the most challenging and rewarding of plastic surgical procedures. Methods of repair have evolved considerably over the past several decades. Presently, all repairs should be soundly founded in the modern principles that have been distilled and clarified from this evolutionary process. This chapter presents a practical synthesis and application of those principles to contemporary bilateral cleft lip repair, in the style of Mulliken.

Keywords: bilateral cleft lip, cleft lip nasal deformity, orofacial cleft

14.1 Introduction

A number of infants born with bilateral cleft lip undergo old-fashioned, often multistaged, procedures, and later, they have to endure sundry revisions throughout childhood and adolescence. Despite the surgeon's best efforts, the stigmata of the repaired cleft lip and nose remain painfully obvious—even at a distance. To the contrary, ... the appearance of a child with repaired bilateral cleft lip should be comparable to, and in many instances surpass, that of a repaired unilateral cleft lip.

— John B. Mulliken

This chapter provides an introduction to the operative treatment of bilateral cleft lip, with or without cleft alveolus and with or without cleft palate. The history of bilateral cleft lip repair is rich, interesting, and lengthy. For those interested, a thorough recounting of this history is available. This chapter focuses on a practical synthesis and application of the principles that have been distilled from this evolutionary process.

14.2 Anatomy, Terminology, and Classification

A history of classification systems for cleft lip or palate summarizes the major types of the bilateral paramedian orofacial cleft deformity, as follows (► Table 14.1):

“Lesser-form” cleft lip is an “umbrella term” that may be further described as minor-form, microform, or mini-microform cleft lip.

Table 14.1 Classification systems for cleft lip and palate

Laterality	Severity	Anatomic involvement	
Bilateral	Complete	Cleft	Lip
	Incomplete		Lip and alveolus (primary palate)
	Lesser form		Lip, alveolus (primary palate), and secondary palate

If the two sides of the bilateral cleft lip are clefted to the same degree (e.g., bilateral complete and bilateral incomplete), the cleft is termed *symmetric*. Contrarily, if the two sides of the bilateral cleft lip are clefted to different degrees (e.g., complete + incomplete and complete + lesser form), then the cleft is termed *asymmetric*. The ontological qualifier “symmetric” considers the severity of only the preforaminal (labial and alveolar) clefts on each side; the term does not encompass other aspects of symmetry that are important to the surgeon, such as the possibility of a deviated premaxilla that is situated closer to one side than the other (► Fig. 14.1).

Premaxillary malpositioning will occur only if the alveolar arch is clefted. In the typical bilateral symmetric cleft lip (e.g., bilateral complete and bilateral incomplete), the premaxilla and vomer usually remain in the midline (*nondeviated*). In the bilateral complete case, the prolabium is separated from the lateral labial elements and the premaxilla is separated from the lateral alveolar processes; this allows unrestrained growth of the vomer and results in anterior *projection* and/or *proclination* of the premaxillary segment. In the bilateral incomplete case, the retention by the partial labial and/or partial alveolar attachment may prevent projection and proclination. In bilateral asymmetric cleft lip (e.g., complete + incomplete), an imbalance of retention forces on one side versus the other results in *deviation* of the premaxilla toward the lesser side.

The morphology of any palatal cleft that may be present will also have an effect on the position of the premaxilla and vomer.

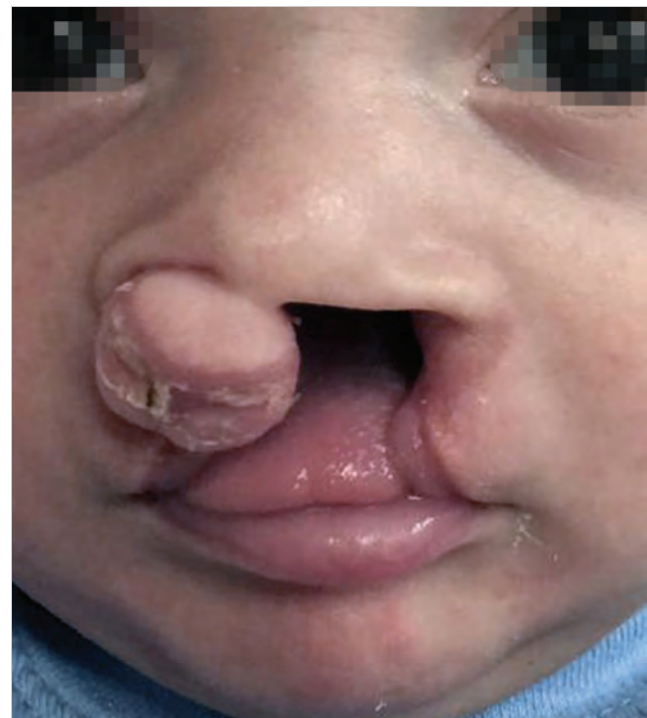


Fig. 14.1 Bilateral complete cleft lip, alveolus, and Veau-IV palate. Although ontologically “symmetric” (because the severity on each side is complete), there is significant rightward deviation of the premaxilla.

In the typical case of bilateral cleft lip with cleft palate, the palatal cleft is morphologically Veau-IV (i.e., a midline cleft of the secondary palate, with exposure of the vomer and bilateral extension through each alveolar process). Of course, it is possible for the palatal cleft to be morphologically different (i.e., Veau-III, Veau-II, and Veau-I), and it is therefore important to describe the morphology of the cleft palate in the complete description of the cleft phenotype. Veau-III cleft palate (unilateral complete cleft through one side of the alveolar arch, with vomerine attachment to the contralateral palatal shelf) results in appreciable deviation of the vomer and premaxilla.

The anatomic details of the bilateral cleft are important for preoperative planning and intraoperative treatment. In the text that follows, we will use a shorthand notation to describe specific phenotypic cases: bCL, bCLA, and bCLAP for bilateral cleft lip; bilateral cleft lip and alveolus; and bilateral cleft lip, alveolus, and palate, respectively. If more phenotypic detail is of significance to the discussion, it will be described in longhand.

14.3 Planning

14.3.1 Principles

The modern approach to primary repair of the bilateral cleft lip and related nasal deformity is founded on several guiding principles:

- Maintain (or establish) symmetry.
- Prepare the projecting premaxilla.
- Anticipate fourth-dimensional changes that occur with growth.
- Construct a full central lip by using lateral labial elements and discard prolabial vermillion.
- Deepen the gingivolabial sulcus by using premaxillary mucosa.
- Establish muscular continuity primarily.
- Address the nasal deformity synchronously.

The first three principles relate to operative planning or preparation, whereas the remaining principles relate to technical details that should be executed skillfully during the operative repair. For thorough exploration of these principles and the history behind them, the reader may refer to Mulliken and Allori and Marcus. Disregard for some of these principles may lead to inferior outcomes and the stigmata of bilateral cleft lip repair (► Fig. 14.2).

14.3.2 Treatment Algorithm

Presentation in Infancy

Ideally, a cleft team will first encounter an infant with cleft lip and/or palate (CL/P) in the neonatal period, perhaps after previously having met the then-expectant parents in prenatal consultation. The purpose of the first encounter is to examine the child, document an accurate phenotype, and begin to formulate and coordinate the treatment plan; to evaluate feeding and nutrition and provide feeding education and support; and to begin the process of preoperative education for the parents.

As described elsewhere in this textbook, protocols for treatment of a child with CL/P may differ from center to center. Generally, the early surgical timeline for a child who presents with the condition in infancy proceeds as follows (► Table 14.2):

For a child with bCL (with intact alveolus and palate), the surgeon may proceed with primary labial repair directly.

For bCLA or bCLAP, the cleft surgeon should first determine whether presurgical orthopedics is indicated and available. If so, this process should be initiated as early as possible in order to achieve maximal effectiveness in a timely fashion, so that operative treatment is not unduly delayed. Presurgical orthopedics is described in detail in a subsequent section of this chapter. In centers where presurgical orthopedics is not available, bilateral labial or nasolabial adhesion may be performed instead. Nasolabial adhesion achieves some of the same effects as presurgical orthopedics on premaxillary repositioning from the natural retrograde pull of the labial elements. Nasolabial



Fig. 14.2 These two patients demonstrate some of the stigmata of bilateral cleft lip repair when some of the principles are not followed. **(a)** Here, a very wide, shield-shaped philtrum is evident. The median tubercle is deficient, and an absence of vermillion is made up for by pseudokeratinized mucosa. There is no white roll under the philtrum. In full smile, the ptotic premaxillary mucosa is evident. The alar base remains wide, and the nasal tip is broad and flat. **(b)** In profile, the columella is short, and the nasal tip lacks projection and definition. Moreover, evident in this photograph are the marked maxillary hypoplasia and retrusion, with negative overjet and class III malocclusion. A late sequela of this is the cleft lip lower lip deformity (CLLLD) that results as it is chronically stretched to ensure oral closure.

Table 14.2 Early surgical time-line for a child with cleft lip or palate

Age	Intervention
0–4 mo of age	Possible presurgical orthopedics
0–4 mo of age	Possible labial or nasolabial adhesion
3–5 mo of age	Primary cheiloplasty (labial repair), with or without gingivoperiosteoplasty and with or without synchronous correction of nasal deformity
10–12 mo of age	Primary palatoplasty

adhesion is described in section “Adhesion Cheiloplasty: Labial Adhesion and Nasolabial Adhesion.”

Nasolabial adhesion may be performed for another reason: it may be utilized to great effect in case of bilateral asymmetric case, in order to preliminarily establish better symmetry before proceeding with lip repair. For example, in case of complete + incomplete bCLA, nasolabial adhesion may be performed *unilaterally* on the complete side, effectively converting the asymmetric phenotype into symmetric bilateral *incomplete* cleft lip and alveolus. The primary labial repair that follows is thereby facilitated in both the design and the execution of the repair. In deciding whether to include nasolabial adhesion for this purpose in his or her treatment protocol, the cleft surgeon must weigh the relative benefit of improved symmetry with the risk and burden of the extra anesthetic and operative procedure.

Late Presentation

For many reasons, including significant comorbidities and international adoption, some children may first present to the cleft team at an older age. At this time, malposition of the premaxilla poses a greater challenge due to the strength and rigidity of the vomer in an older child; that is, although the vomer of the neonate may be “buckled” by presurgical orthopedic techniques or by nasolabial adhesion, this is no longer possible in the older child.

If the premaxillary projection or proclination in the older child is minor, it may be possible to perform preliminary adhesion or to proceed directly to labial repair, with minimal tension on the labial repair. On the other hand, if the premaxilla is severely projecting or proclined, this precludes a safe and effective labial repair, because the gap that must be spanned by the cutaneous and muscular closure is too great and will be under excessive tension. In order to repair the lip over this gap, the surgeon is forced to design an unaesthetically wide philtral flap; moreover, the size of the gap makes continuous muscular approximation difficult and gingivoperiosteoplasty (GPP) impossible.

In addition to the challenges of premaxillary projection or proclination, premaxillary deviation is also difficult to correct in the older child. This deviation results in an asymmetry that cannot be compensated for merely by alteration of the design of the repair.

In such cases, the skeletal malposition requires a skeletal correction: premaxillary osteotomy and setback. This procedure is described in section 14.4.3, “Premaxillary Osteotomy and Setback”. Importantly, premaxillary setback must be done as a

preliminary procedure and should never be combined with primary cheiloplasty. In premaxillary osteotomy, the division of the vomer disrupts the posterior blood supply to the premaxilla, leaving the premaxilla totally dependent on anterior blood supply from the prolabium. If the philtral flap is dissected and raised, this will also eliminate the anterior blood supply, risking avascular necrosis of the premaxilla. Consequently, if premaxillary osteotomy and setback are necessary, they must be done 3 to 4 months before the lip repair. It is safe to combine premaxillary osteotomy and setback with either nasolabial adhesion (in case of bCLA) or palatoplasty with GPP (in case of bCLAP). These procedures improve the fixation and support of the retropositioned premaxilla, while respecting its prolabial blood supply. In additionally, in case of an older child with bCLAP, performing palatoplasty at this time efficiently combines two procedures that this child needs under one anesthetic. The definitive labial repair (with inherent elevation of the philtral flap) may be safely performed approximately 3 to 4 months following the premaxillary setback.

14.4 Preparation

14.4.1 Presurgical Orthopedics

Presurgical orthopedics refers to the remodeling of the dentofacial skeleton by way of orthodontic techniques. Generally, this process may be either *active* or *passive*. Active processes use mechanical gears and levers to exert force on the dentofacial skeleton in order to remodel it quickly. An example of an active device is the Georgiade–Latham device, or pin-retained elastic chain premaxillary retraction (ECPR) device. In contrast, a passive device gradually remodels the dentofacial skeleton in response to statically sustained extrinsic forces that are increased at periodic intervals. Examples include lip taping, palatal splints, and nasoalveolar molding (NAM). Both approaches require the participation and expertise of craniofacial orthodontists. The various techniques of presurgical orthopedics have been debated fiercely. The merits and demerits of the most common methods are worthy of review and have been summarized recently in an excellent point/counterpoint article.

Both the Georgiade–Latham’s device and NAM are effective at widening a collapsed maxillary arch and at retropositioning the projecting and retroclining the proclined premaxilla. As an active technology, the Georgiade–Latham’s device is quite rapid, with treatment completed over the course of 3 to 6 weeks. In contrast, NAM requires 3 to 4 months. The Georgiade–Latham’s device requires a separate anesthetic for initial device placement, but it may be activated by parents (by turning a screw) at home, with periodic clinical check-ups. Although NAM does not require an anesthetic at any point during treatment, it does require weekly clinical visits in order to modify the appliance. One significant advantage that NAM has over the Georgiade–Latham’s device is that midway through treatment, outrigger nasal prongs are added to begin to round out the flattened nasal ala. Although many single-center, long-term case series have been published that demonstrate the efficacy of this technique, there remains considerable debate in the literature regarding its widespread effectiveness.

14.4.2 Adhesion Cheiloplasty: Labial Adhesion and Nasolabial Adhesion

In circumstances where presurgical orthopedics are not available and where premaxillary protrusion, proclination, and deviation are significant, the surgeon may opt to perform nasolabial adhesion in order to indirectly achieve retropositioning and centralization of the premaxilla. The basic principle behind adhesion is to connect each side of the cleft, utilizing areas of the cleft margin that will be discarded in the definitive repair. To perform adhesion, it is advised that the surgeon first draw the markings for the definitive cheiloplasty, which will be performed later. The adhesion should be performed by utilizing the tissues of the prolabium and lateral lip elements that will be discarded in the future repair. In this fashion, any scar that forms during the adhesion would be discarded. Importantly, dissection is minimized in order to prevent excessive scarring, which would complicate the future repair. *Labial* adhesion connects the cutaneous lip in an “open-book” fashion, whereas *nasolabial* adhesion extends this superiorly to preliminarily close the nasal sill, utilizing the columellar flaps. In either case, there is no muscular dissection, and the prolabial skin should not be elevated off the premaxilla. The technique for adhesion is described further in this textbook in the chapter on unilateral cleft lip repair.

14.4.3 Premaxillary Osteotomy and Setback

The method of premaxillary osteotomy and setback directly achieves retropositioning and centralization of the premaxilla. It is usually reserved for the older child who is no longer a candidate for presurgical orthopedics or nasolabial adhesion.

To perform the technique, the patient is placed on the operating room table in supine position, with a shoulder roll to provide gentle neck extension. A Dingman's oral retractor is placed to provide adequate exposure of the palate. In case of bCLA, the secondary palate will be intact, but the vomer will be visible through a defect in the primary (preforaminal) palate. In case of bCLAP, the cleft palate will expose the vomer in its entirety.

For bCLA, the intention is to retroposition the premaxilla, close the nasal mucosa, close the oral mucosa, and perform GPP. After infiltration of local anesthesia with epinephrine, a midline incision is made on the mucosa overlying the exposed region of the vomer. A Cottle's periosteal elevator is used to elevate the mucoperiosteal vomerine flaps, exposing the underlying bone and the premaxillary–vomerine suture. Next, incisions should be made at the margin of the primary palatal cleft, and the Cottle's elevator should be used to separate the nasal mucosal lining from the oral mucosa, extending this dissection posteriorly above and below the palatal shelves. Similarly, incision should be made in the mucosa on the posterior aspect of the premaxilla, and this should be gently reflected to provide flaps to which the nasal and oral mucosa may be sutured. The osteotomy is then performed immediately posterior to the premaxillary–vomerine suture, using a rongeur. The amount of bone that must be removed is dependent on how much it needs to be moved backward, and the shape of bone removal (whether rectangular or trapezoidal) is dependent on whether the

premaxilla needs to be rotated as well. In actuality, bone is removed incrementally until the proper position is achieved. It is critical not to remove too much bone because direct bone-to-bone contact is required for osseous union. Rigid fixation is not required. Some surgeons pass a Kirschner's wire through the premaxilla into the vomer, which provides temporary semirigid fixation. It is the practice of the author to secure the bone with one 2–0 Vicryl suture, which prevents undue movement during the immediate postoperative period. Some surgeons do not fixate the bone at all and just rely on the mucosal closure and GPP to tether the premaxilla in its new position. Next, the nasal mucosa should be closed by using 5–0 Vicryl or chromic suture, and the oral mucosa should be approximated by using 4–0 Vicryl. The GPP should then be completed in the typical fashion.

For bCLAP, the intention is to secure the retropositioned premaxilla by way of the palatal repair and GPP. In this case, the procedure starts by designing the typical two-flap palatoplasty for repair of the Veau-IV cleft palate, as described in detail elsewhere in this textbook. After elevation of the oral mucoperiosteal flaps and elevation of the nasal mucosal lining from the palatal shelves, the vomerine flaps are elevated from the underlying vomer. Prior to closure of the nasal floor, the premaxillary osteotomy is performed, and the premaxilla is retropositioned, as described earlier. The vomerine flaps are then used to close the nasal floor on each side, and this nasal mucosal closure is completed anteriorly at the premaxilla. Gingivoperiosteoplasty is then completed to close the alveolar clefts and further stabilize the new position of the premaxilla. The palatal repair is then completed in the typical fashion (► Fig. 14.3).

For infants, the author recommends the placement of a temporary petroleum gauze palatal splint for 1 day. It is removed before discharge from the hospital.

For an older child with deciduous dentition, it is recommended that a custom-made dental splint be made that will protect the premaxilla in its new position. This is more important to protect the older child (who may injure himself or herself during play) than for the purpose of semirigid fixation.

14.5 Primary Repair of Bilateral Cleft Lip

Although several techniques for bilateral cleft lip repair have been described, the Mulliken's technique is described as follows.

14.5.1 Marking

A wide double hook is placed in the nostrils to gently elevate the nasal tip and establish symmetry, if needed. The standard anthropometric landmarks should be marked:

- Nasion (n).
- Endocanthion (en).
- Pronasale (prn).
- Subnasale (sn).
- Ala nasi (al).
- Subalaris (sbal).
- Labiale superius (ls).
- Crista philtri superioris (cphs).

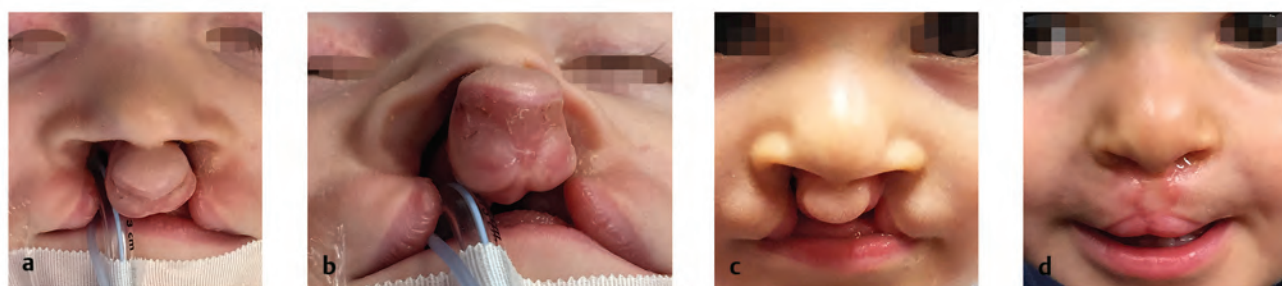


Fig. 14.3 A 10-month-old infant with bilateral complete cleft lip, alveolus, and Veau-IV palate, presented too late for presurgical orthopedics, requiring premaxillary osteotomy and setback. (a,b) The premaxilla is significantly projecting and moderately proclined. (c) Premaxillary osteotomy and setback were performed in combination with palatoplasty. Gingivoperiosteoplasty could not be completed due to the width of the alveolar cleft. (d) The cleft lip repair is completed 3 months later. Healing in this case was complicated by scar hypertrophy, especially lateral to the left side of the philtral flap.

- Crista philtri inferioris (cphi).
- Stomion (sto).
- Chelion (ch).

When taking measurements for symmetry, sn should serve as the main reference for prolabial markings and sbal for lateral lip markings. In addition to the above landmarks, the vermilion-mucosal junction (“red line”) of the lip is indicated with a dotted line.

On the prolabium, the philtral flap is drawn. For a child younger than 1 year, the philtral flap should be designed to be 2-mm wide superiorly (cphs–cphs), 4-mm wide inferiorly (cphi–cphi), and 6 to 7 mm vertically (sn–ls). In complete cleft lip, the total available height of the prolabium is often 6 to 7 mm. In incomplete cleft lip, the prolabium may actually be longer, and in this case, the excess prolabial height will not be included in the design of the philtral flap. The sides of the philtral flap are concave. The philtral flap is bordered on each side by 2 to 3 mm “flanking flaps” that will be deepithelialized. These flanking flaps provide extra bulk for the philtral ridge and also improve the vascular supply to the philtral flap. At the top of the prolabium, horizontal lines are drawn to indicate the C-flaps that are preserved and utilized in the closure of the nasal floor.

On the lateral labial elements, Noordhoff’s point is marked at the medial-most location of the lip, where a distinct white roll is present and where the height of vermilion is maximal. This point may be adjusted slightly to ensure that there are 3 mm of white roll medial to the proposed location of the cphi point of the Cupid’s bow. It is important not to place cphi too medially, where the white roll has become indistinct or where the vermilion has begun to taper. In addition, it is important not to place the point too laterally, as this will result in a tight closure and possibly microstomia. Generally, the cphi–ch distance should be greater than 15 mm on each side. The lateral labial cutaneous advancement flaps are drawn as large as possible, given they will be trimmed to fit later in the procedure. In marking these flaps, the white roll should remain on the side of the vermilion. At its superior aspect, the marking for the advancement flap curves around the alar base up to, or just beyond, sbal (► Fig. 14.4).

Dilute anesthetic with epinephrine should be infiltrated. Key landmarks are tattooed with methylene blue, and a scalpel may be used to score the markings.

14.5.2 Operative Technique

The general operative sequence is shown as follows; however, in actuality, some maneuvers are interdependent:

1. Philtral flap and lateral labial dissection.
2. Mucosal flap for deepening of the gingivolabial sulcus.
3. Gingivoperiosteoplasty.
4. Closure of the nasal floor.
5. Labial closure.
6. Nasal correction.
7. Skin closure.

Labial Dissection

The operation begins with dissection of the philtral flap. The incision is made with a #69a mini blade or #15c scalpel. Great care should be taken in the deepithelialization of the flanking flaps, such that the dermis is preserved and remains attached to the philtral flap. The remaining prolabial skin is removed and discarded. The philtral flap (en bloc with the flanking flaps) is then elevated superiorly, and sharp scissors are used to complete the dissection in the subcutaneous plane to the level of the anterior nasal spine.

The lateral labial flaps are incised as marked, and the vermilion-mucosal flaps are separated sharply from the musculocutaneous portion of the lip. Near the alar base, sharp dissection is performed submuscularly to the level of the maxilla and then continued laterally in the supraperiosteal plane. The attachment of the alar base to the piriform rim is released. A Tessier’s elevator is used to complete this supraperiosteal mobilization of the soft tissue of the cheeks as far laterally as the malar eminences. This dissection is important for reducing the tension on the lip repair.

The cutaneous advancement flap is secured with fine double hooks, and scissors are used to sharply dissect the orbicularis oris muscle from the overlying skin and the underlying oral mucosa.



Fig. 14.4 (a) A newborn with bilateral complete cleft lip, alveolus, and Veau-IV cleft palate, who underwent (b) nasoalveolar molding in preparation for lip repair. (c,d) Following NAM, there was minor residual deviation of the premaxilla, which was compensated for in the design (e) of the repair. (f,g) The final two photographs show the result at the time of suture removal.

Mucosal Flap and Gingivoperiosteoplasty

Next, the subcutaneous or submucosal tissue overlying the premaxilla is sharply removed. The thin strip of prolabial vermillion is removed and discarded. The premaxillary mucosa is elevated to form an inferiorly based mucosal flap, which is then pulled upward and secured to the premaxillary periosteum at the level of the anterior nasal spine by using Vicryl or polydioxanone (PDS) suture. This flap forms the posterior wall of the new gingivolabial sulcus. If the mucosal flap is not lifted superiorly, the gingivolabial sulcus will be very shallow and premaxillary mucosa will hang down and create the appearance of a “double lip” when the child smiles.

Gingivoperiosteoplasty is completed at the same time as the mucosal flap creation. The gingiva and periosteum of the lateral alveolar segments are incised vertically. The gingivoperiosteal flaps are secured by using 6-0 or 5-0 chromic suture, beginning posteriorly and proceeding anteriorly.

Closure of the Nasal Vestibule and Nasal Sill

Medial nasal mucosal flaps are elevated from the premaxilla, and corresponding lateral nasal mucosal flaps are elevated from the inferior turbinates. Closure of the vestibular mucosa proceeds from posterior toward anterior side.

Closure of the cutaneous nasal sill is completed later in the procedure, after beginning the labial closure and repositioning the nasal cartilages. At that time, the alar base is transposed medially and sutured to the prolabial C-flaps by using 7-0 Vicryl suture for the dermis and 7-0 chromic suture for the skin. The columellar flaps may require trimming to achieve the ideal alar base width. An alar base cinch suture (5-0 Prolene, passed through the dermis of each alar base near sbal and

through periosteum of anterior nasal spine) is used to precisely control the alar base width. Myrtilform sutures (4-0 PDS suture placed in maxillary periosteum just medial to each canine fossa and passed through the midpoint of each nasal sill) may be used to slightly depress the nasal sill and floor.

Labial Closure

After completion of the GPP and mucosal flaps, the lateral labial mucosa is next to be closed. An inferolateral back cut may be necessary for tension-free advancement of the closure. A 5-0 chromic suture may be used for the closure. In performing this step, it is extremely important to apply firm traction medially during the closure in order to adequately advance the flaps.

Next, orbicularis oris muscle is approximated in the midline by using 4-0 Vicryl or PDS suture in horizontal mattress fashion. The superior aspect of the orbicularis oris muscle is secured to the periosteum of the anterior nasal spine by using 4-0 Prolene suture to prevent it from sliding inferiorly with facial animation.

The vermillion flaps are then used for creation of the median tubercle. The previously tattooed cphi points are identified on each side, and a mark is made 3 mm medial to each. A 7-0 Vicryl suture and a 7-0 chromic suture are used to approximate these marks to create the new ls point, into which the point of the philtral flap will later be inset. Vermilion closure proceeds down the midline to create the median tubercle. Excess vermillion or mucosa may be deepithelialized/demucosalized, with the submucosa used to augment the volume of the median tubercle.

Nasal Correction

As the cleft lip is repaired and the alar base width is narrowed, the cleft lip nasal deformity (CLND) often worsens drastically.

Therefore, synchronous correction of the CLND has been advocated. Several approaches have been described, including use of percutaneous transfixion sutures and semiopen approach. In the semiopen approach, the medial crura of the lower lateral cartilages (LLCs) are mobilized beneath the C-flap through the open labial incisions. Bilateral alar rim incisions or marginal incisions are used to expose the inferior aspect of the LLCs. Great care must be taken not to injure the cartilages during this process. The LLCs are exposed and cleaned of fibrofatty tissues on their superficial and medial surfaces. Similarly, the upper lateral cartilages (ULCs) are exposed. A cotton-tip applicator may be placed into the nasal vestibule, and gentle upward traction is used to visualize the location of the genua to be created in each LLC. Medial to each genu, two interdomal 5–0 PDS sutures are placed between the domes of each LLC but are left untied. Lateral to each genu, two intercartilaginous sutures are used to re-create the interface between the LLC and the ULC (“scroll region”). With the cotton-tip applicator assisting with proper positioning of the cartilages in relation to each other, the sutures are sequentially tied. The skin is then closed by using 7–0 chromic suture. Any redundant domal skin can be carefully excised in crescentic fashion to improve the contour of the alar rim. At this point, any preexisting vestibular web may be accentuated. This can be corrected through lenticular excision of mucosa and closure.

Skin Closure

To complete the procedure, the tip of the philtral flap is inset to meet the ls point of Cupid’s bow. A 7–0 Vicryl suture is used to reapproximate dermis, and a 7–0 nylon suture is used for skin closure. The lateral labial cutaneous advancement flaps are trimmed superiorly to conform to the shape of the nasal sill. Closure is completed in two layers, with 7–0 Vicryl and 7–0 chromic sutures.

14.5.3 Postoperative Care

At the completion of the procedure, the face is washed and dried. Bacitracin ointment may be applied to the incision line. It is the author’s practice to place temporary vented stents (Xeroform wrapped around 19-gauge butterfly tubing and cut to 1.25-cm length) in each nostril. These are removed before the patient is discharged from the hospital. Some surgeons tape a Logan’s bow to the cheeks to support an iced saline gauze over the incision line to reduce bloody crusting in the perioperative period.

The patient should be observed overnight. The parents should be educated on careful feeding techniques and on lip hygiene. Any bloody crusting should be gently cleaned with half-strength hydrogen peroxide and saline, and antibiotic ointment should be reapplied. It is important that the parents feel

comfortable with suture-line care and are well aware of how to keep the nostrils clean.

One week postoperatively, sutures are removed under general anesthesia by using mask induction and insufflation. A Steri-Strip is placed from side to side to help protect the healing incisions. This Steri-Strip is replaced as needed over the course of the next 2 to 4 weeks.

Further postoperative follow-up occurs at 1 month and 3 months. At 1 month, the parents are instructed on how to perform scar massage and are advised to apply sunblock (SPF ≥ 50) in the morning and skin lotion at night. At times, a thickened scar that starts to cause contracture requires intralesional injection of Kenalog (triamcinolone). This is performed in the clinic and may be repeated every 4 to 6 weeks up to three times. Scar massage should continue throughout this period until the scar softens and flattens.

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15 Cleft Palate

Ingrid Ganske and Carolyn R. Rogers-Vizena

Summary

Cleft palate is a congenital craniofacial anomaly that affects feeding, hearing, and speech. The hard palate gives structural support, and the soft palate provides velopharyngeal competence. The aim of cleft palate repair is to close the oronasal fistula and to construct a dynamic velar sling, thereby establishing oronasal competence and enabling control of airflow for intelligible speech. With this goal in mind, palate anatomy, perioperative concerns, surgical technique, postoperative care, and palatoplasty complications are described in this chapter.

Keywords: cleft palate, cleft palate repair, palatoplasty, Robin sequence, velopharyngeal, speech surgery

15.1 Introduction

Detailed understanding of palate development, normal and cleft palate anatomy, and epidemiology of clefting is essential to the comprehensive care of children with cleft palate.

15.1.1 Embryology

Palatal processes form from the maxillary prominences during the seventh week postconception. These vertically oriented shelves elevate to a horizontal position by the ninth week postconception. They meet with each other to form the secondary palate and also fuse with the primary palate. Failure of elevation and fusion of these shelves result in a cleft palate.

15.1.2 Anatomy

Normal Palate Anatomy

The incisive foramen is located at the midline, posterior to the alveolus, at the junction of the primary and secondary palate.

Structures anterior to the incisive foramen are considered the primary palate, and structures posterior to the incisive foramen are considered the secondary palate. The secondary palate comprises both hard and soft palate. The hard palate is the growth center for the maxilla and is formed by paired palatine processes of the maxilla and palatine bones, articulating with the vomer. The soft palate is a muscular sling formed by the paired *levator veli palatini*, *palatopharyngeus*, *palatoglossus*, and uvular muscles (innervated by the pharyngeal branch of the vagus nerves) and is reinforced by the broad aponeurosis of the *tensor veli palatini* (innervated by the medial pterygoid branch of the mandibular nerves). These muscles work with the superior constrictor to form the velopharyngeal sphincter.

Palatal and velopharyngeal blood supply comes from branches of the paired internal maxillary arteries, recurrent pharyngeal arteries, ascending pharyngeal arteries, and ascending palatine arteries. The soft palate and velopharyngeal sphincter muscles have a rich blood supply from branches of these arteries. The greater palatine branches of the internal maxillary artery provide the major blood supply to mucoperiosteum of the hard palate (► Fig. 15.1).

Cleft Palate Anatomy

Isolated cleft palate involves secondary palatal structures. Maxillary and palatine bones are separated at the midline, resulting in an oronasal fistula. The *tensor veli palatini* aponeurosis, *levator veli palatini* muscles, and *palatopharyngeus* muscles are disoriented parallel to the cleft, inserting on the posterior hard palate (► Fig. 15.1). As a result, function of the velopharyngeal sphincter is compromised. These muscles normally participate in ventilation and pressure equalization of the middle ear. Altered anatomy of these muscles results in Eustachian tube dysfunction, frequently causing chronic middle ear effusion, hearing loss, and otitis media.

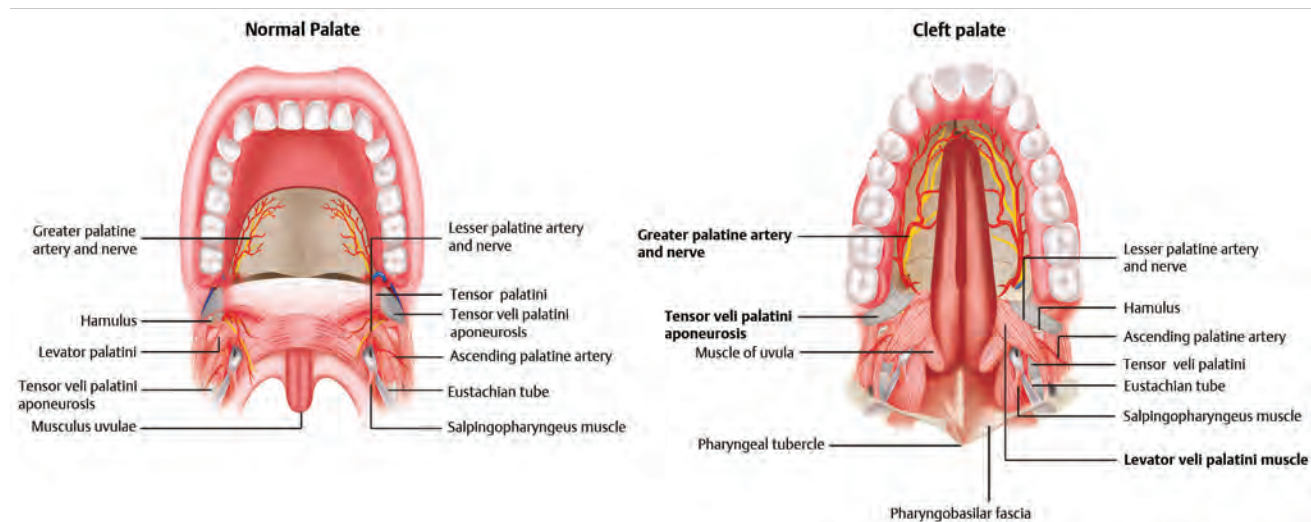


Fig. 15.1 Palate anatomy.

In cleft lip and palate, the defect extends along the junction of the primary and secondary palate, resulting in an alveolar gap. When the alveolus is cleft, lateral incisors are frequently absent and other dental anomalies, such as supernumerary teeth, are common. The palatal component of orofacial clefts is often described using the Veau classification (► Fig. 15.2).

Submucous cleft palate is a lesser form of cleft palate involving muscular diastasis of the soft palate, with intact palatal mucosa. The diastasis may be visualized as a bluish “zona pellucida,” or furrow, in the soft palate. Other findings may include a bifid uvula and/or a notch in the posterior hard palate (► Fig. 15.3). These three findings constitute Calnan’s triad. This type of cleft is often diagnosed during evaluation of abnormal speech.

15.1.3 Epidemiology

Traditionally, cleft lip with or without cleft palate (CL/P) and isolated cleft palate (CP), also known as cleft palate only, have been considered separate etiopathogenetic entities; however, emerging evidence suggests that the delineation is less distinct. In the United States, recent data from the National Institute of Dental and Craniofacial Research estimate the incidence of CL/P to be approximately 1 in 940 live births. The condition is twice

as common in male patients compared with female patients and has ethnic heterogeneity, with Asians and Hispanics demonstrating the highest incidence, Caucasians demonstrating intermediate incidence, and Africans manifesting the lowest incidence. Estimated incidence of CP is approximately 1 in 1,570 live births. The condition is twice as common in female patients and does not have the same ethnic variation that CL/P has.

Clefts of the lip and/or palate result from a combination of genetic and environmental factors. Environmental factors, including maternal smoking, folate deficiency, alcohol consumption, advanced maternal age, and retinoid use, are associated with increased risk. Mutations in multiple candidate genes, such as *IRF6*, *Wnt* signaling, *MSX1*, and *BMP* signaling factors, have also been associated with clefting. Cleft lip or palate is infrequently (10–14%) associated with a recognized syndrome, but 29 to 45% of patients with a cleft lip have associated anomalies, suggesting possible underdetection. Among the syndromes associated with CL/P are Van der Woude syndrome, oral-facial-digital syndrome, branchio-oculo-facial syndrome, and Opitz G/BBB syndrome. However, isolated CP is often (12–54%) associated with an underlying syndrome, and as many as 47 to 72% of patients have associated anomalies. Among more than 250 syndromes associated with CP are Stickler’s syndrome,

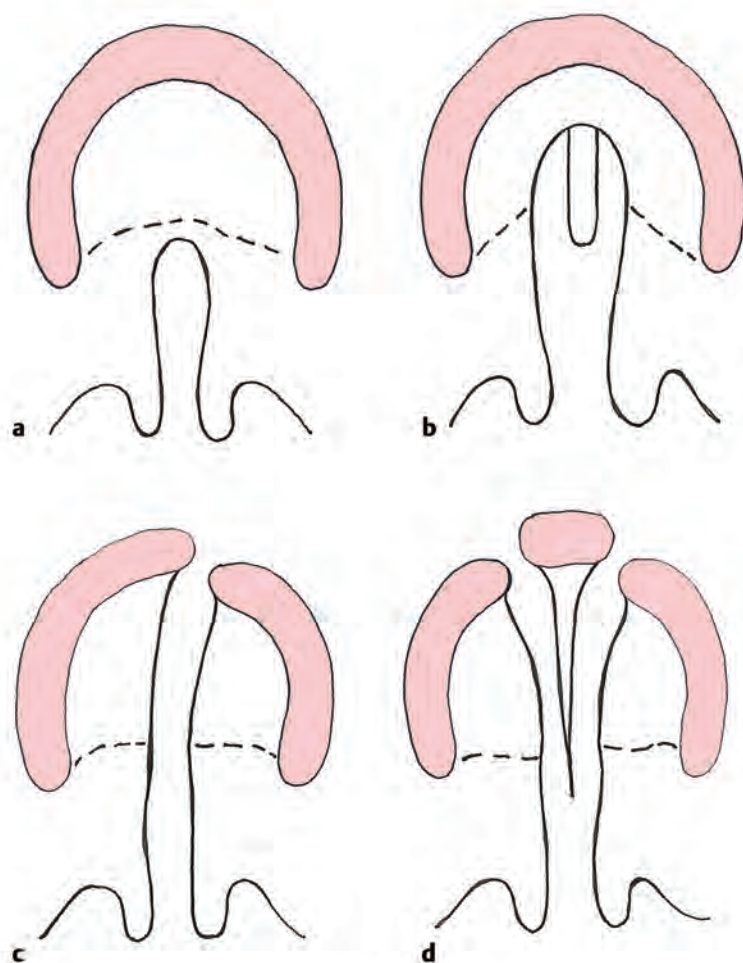


Fig. 15.2 Veau classification of cleft palate. (a) Veau I: Soft palate cleft only. (b) Veau II: Soft and hard palate cleft, posterior to the incisive foramen. (c) Veau III: Unilateral cleft lip and palate, through alveolus, hard, and soft palate. (d) Veau IV: Bilateral cleft lip, through the alveolus bilaterally, hard palate, and soft palate.

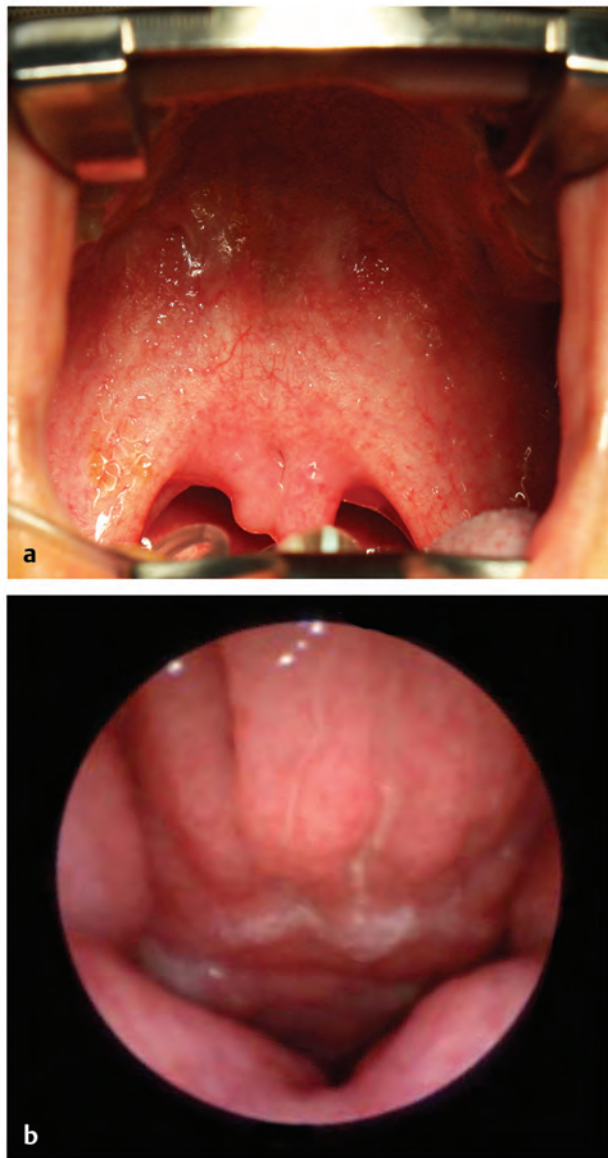


Fig. 15.3 Submucous cleft palate. (a) Intraoral view of an overt submucous cleft palate demonstrating a bifid uvula and dark-colored midline depression caused by muscular diastasis. (b) Nasal endoscopic view of the same patient. With speech, a deep furrow is seen on the nasal surface of the soft palate, also indicating muscular diastasis.

22q11 deletion spectrum, Cornelia de Lange syndrome, Emanuel syndrome, trisomy 13, Treacher Collins syndrome, and Apert's syndrome.

15.2 Diagnosis

Clefts of the lip and palate are increasingly diagnosed prenatally. Cleft lip is often noted on routine prenatal examination, because the primary palate is well visualized on sonography (► Fig. 15.4a). Isolated CP is rarely diagnosed with sonography but may be incidentally detected with fetal magnetic resonance imaging (MRI) (► Fig. 15.4b) performed as a part of the evaluation of other congenital anomalies. In most cases, diagnosis of

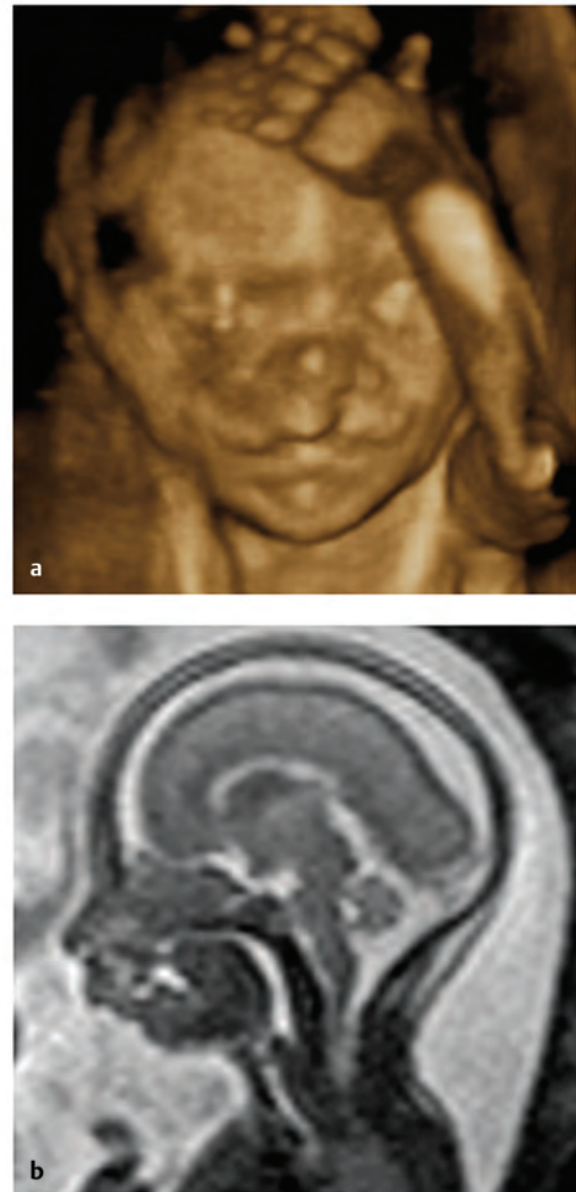


Fig. 15.4 Prenatal diagnosis of cleft lip and palate. (a) Fetal three-dimensional sonography demonstrating a left unilateral incomplete cleft lip. (b) Fetal magnetic resonance image demonstrating a Veau II cleft palate. The fetal tongue rides high into the nasal cavity due to the cleft hard and soft palate.

CP is made by physical examination postnatally. As soon as the diagnosis is made, the expectant mother or the newborn should be referred to a cleft team for evaluation and counseling.

15.3 Nonoperative Management

The concept of team care is one of the most important characteristics of modern cleft care. The American Cleft Palate-Craniofacial Association and Cleft Palate Foundation set standards for designated cleft centers. A cleft team must include a speech and language pathologist, a surgeon trained in cleft lip and palate

repair, and an orthodontist. In addition, to best serve the unique needs of children with a cleft lip or palate, the team should have access to practitioners in psychology, social work, psychiatry, audiology, genetics, general and pediatric dentistry, otolaryngology, and pediatrics or primary care.

Infants with CP often have feeding difficulty early in life. When a cleft palate is present, infants are not able to generate suction due to the large oronasal fistula. Feeding instruction by a speech and language pathologist or a nurse skilled in assessing and feeding infants with cleft palate is initiated as soon as the diagnosis is made, either prenatally or shortly after birth. Infants with a cleft palate are best fed in semi-upright position, using cleft feeders. No one feeding system is suited to every infant. The two most commonly used at our center are Medela SpecialNeeds (Haberman) Feeder and Dr. Brown's Specialty Feeding System. Other popular cleft bottles include the Mead Johnson's cleft lip/palate nurser and Pigeon cleft palate nipple. The early management of infants with cleft palate is focused on feeding, weight gain, and nutrition.

15.4 Operative Treatment

15.4.1 Timing of Surgery

Timing for palatoplasty varies widely between cleft centers. Advocates for early repair cite improved speech development. In one study, intervention at 6 months of age led to better speech outcomes, with fewer compensatory articulations than repair at 12 months of age. Another study correlated increasing age at repair to velopharyngeal insufficiency (VPI) requiring corrective surgery, with a major increase when intervention was performed at 13 months of age or later. Because the maxilla is the center of midfacial growth, advocates for later palatoplasty are concerned that early palate manipulation substantially inhibits midfacial growth. The surgeon must balance the potential growth restriction associated with early repair with the poorer speech outcomes associated with late repair. For this reason, a few centers perform hard and soft palate repair separately. However, most surgeons (85%) perform cleft palate repair in a single stage between 6 and 12 months of age. At our center, palatoplasty is typically performed between 9 and 11 months of age.

Palate repair may be delayed if other medical issues are present and not optimized, such as poor weight gain, cardiac anomalies, and airway issues. Robin sequence (also called Pierre Robin sequence or "syndrome"), defined by the triad of microretrognathia, glossoptosis, and airway obstruction, deserves special mention. Isolated CP is found in 69 to 90% of patients with Robin sequence. Robin sequence is often associated with an underlying diagnosis, most commonly Stickler syndrome, which is present in one third to half of patients. Palatoplasty in this subset should be approached cautiously. Airway problems in the early postoperative period have been reported. Repair should be delayed if the airway is not clearly secure.

15.4.2 Palatoplasty

The major goals of modern cleft palate repair are oronasal fistula closure and speech optimization through proper musculature positioning. To close the hard palate, nasal lining is

elevated from the vomer medially and from the maxilla and palatine bones laterally and is approximated to restore nasopharyngeal lining. Palatal mucosa is elevated in a subperiosteal plane and is medialized. Von Langenbeck described elevating the palatal mucosa as bipedicle flaps. The "two-flap palatoplasty," advocated by Bardach, employs a single pedicle based on the greater palatine arteries. In both variations of hard palate repair, a thin area of exposed palatal bone is left laterally to heal by secondary intention. Soft palate repair is performed in three layers: nasal mucosa, velar musculature, and palatal mucosa. In a straight-line repair, the velar musculature is released from the posterior hard palate, rotated posteromedially, and sutured at the midline to create a muscular sling, the "intravelar veloplasty." This technique is widely applicable to all variations of cleft palate, regardless of width. An alternative is the Furlow double-opposing Z-plasty, which both restores the velar sling and lengthens the soft palate. At our institution, the Furlow procedure is used for the initial repair of submucous cleft palate and isolated clefts of the soft palate, as well as for secondary lengthening procedures for VPI. These two techniques are described in detail as follows.

Two-Flap Palatoplasty with Intravelar Veloplasty

Preparation and Markings

The patient is positioned with neck in extension. A Dingman mouth gag maintains retraction. The two-flap palatoplasty is marked (► Fig. 15.5a, b). Local anesthetic with epinephrine is injected into hard and soft palate mucosa for hemostasis and analgesia.

Operative Technique

The soft palate mucosa is incised with a #15 blade along the junction of the nasal and oral mucosa, extending from hard palate to uvula. A #12 blade may be used to complete the uvular incision. The hard palate mucosa is incised laterally, at the border of the alveolus, and medially, at the junction of nasal and oral mucosa. Hard palate mucoperiosteal flaps are elevated in the subperiosteal plane (► Fig. 15.5c), taking care to preserve the greater palatine arteries and dissecting circumferentially around the vessels (► Fig. 15.5d). For wide clefts, the arteries may be cautiously skeletonized, and/or ostectomy at the greater palatine foramen allows further medialization. An incision is made down the vomer, and a periosteal elevator is used to elevate just enough mucoperiosteum to close the nasal lining (► Fig. 15.5e). Attention is focused again to the soft palate. The aggregates of the levator palatini and palatopharyngeus muscles are sharply dissected, and their attachments to the posterior hard palate are released (► Fig. 15.5f).

Closure

Nasal lining is closed using 5-0 chromic suture and 4-0 Vicryl suture, beginning anteriorly and progressing posteriorly (► Fig. 15.5g). Care is taken to evert the flaps and to avoid tearing fragile tissue. The palatal muscles are approximated with 4-0 polydioxanone (PDS) suture in a figure-of-eight fashion. Palatal mucoperiosteum and soft palate mucosa are closed

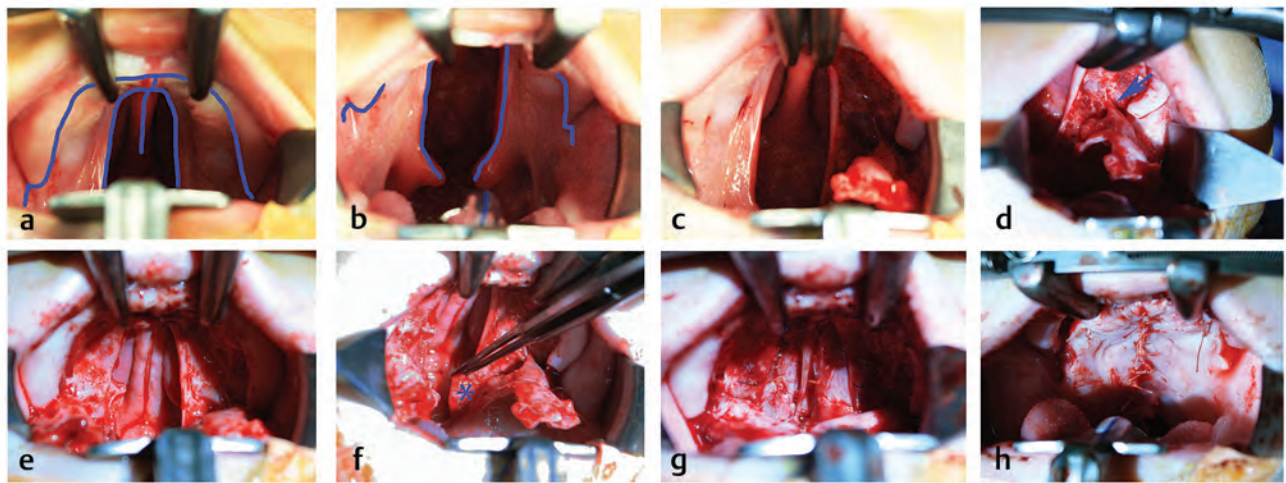


Fig. 15.5 Two-flap palatoplasty (Bardach). (a) Palatal gingivoperiosteal flaps are marked out (blue lines) on either side, based on the greater palatine arteries. The lateral marking is made at the junction of the alveolar gingiva and the palatal mucosa, and the medial marking is made at the junction of the palatal and nasal mucosa. (b) The nasal–oral junction incision extends down the soft palate to the tips of the uvula. (c) The gingivoperiosteal flap is elevated in a subperiosteal plane. (d) Circumferential dissection is performed around the greater palatine vessels (blue arrow). (e) Hard palate flaps are elevated bilaterally. Next, vomerine mucosa is incised and elevated medially, and then, nasal lining is elevated laterally. (f) The velar muscles (*) are dissected and posteromedially rotated. (g) Bilateral vomer and nasal lining flaps are approximated, followed by muscular repair. (h) The oral lining is closed, and gingivoperiosteal flaps are tacked into place.

using 4–0 Vicryl simple interrupted sutures, beginning posteriorly at the uvula and progressing anteriorly. Mattress sutures may be used when eversion is inadequate. The hard palate mucoperiosteal flaps are loosely approximated to the alveolar gingiva anteriorly (► Fig. 15.5h). Occasionally, a suture can be placed through the soft bone of the hard palate to secure the oral layer anteriorly, if needed.

Furlow's Palatoplasty

Preparation and Markings

The oral mucosa is marked with a Z-plasty. The medial cleft margin is marked. The patient's left side is conventionally the musculomucosal posteriorly rotating oral flap, and the right side is the mucosal anteriorly rotating flap (► Fig. 15.6a). The nasal lining is incised in mirror image, with the musculomucosal posteriorly rotating flap on the right and the mucosal anteriorly rotating flap on the left. The incision on the left extends from the junction of the hard and soft palate medially toward the hamulus laterally, at an angle of ~ 60 degrees. The right-sided oral mucosal flap is marked by joining the hamulus laterally to the right side of the uvular base medially, at an angle of ~ 70 to 80 degrees. We have been most successful waiting to mark the exact placement of this incision until after the left side has been cut and designing the flap based on how the tissue best lies in a minimal-tension position. Local anesthetic with epinephrine is injected into the hard and soft palate.

Operative Technique

The left-sided cleft margin is incised with a #15 blade, followed by the posteriorly rotating left oral musculomucosal flap. The velar muscles are elevated with the flaps that rotate posteriorly, oral on the left and nasal on the right, and ultimately, they overlap to create a muscular sling. The levator is sharply dissected

off the posterior edge of the hard palate, and the musculomucosal flap is dissected off the nasal mucosal lining, leaving adequate submucosa on that flap to preserve vascularity. The flap is mobilized to the point of being inset with minimal tension (► Fig. 15.6b). The right-sided cleft margin and lateral limb are incised. The anteriorly based mucosal flap is dissected in a submucosal plane, continuing anteriorly to the posterior edge of the hard palate. Dissection along the edge of the hard palate is performed with a Kramer elevator, freeing the nasal-sided mucosa in this region. Next, the nasal flaps are incised in a mirror image to the oral flaps, musculomucosal flap on the right and mucosal flap on the left (► Fig. 15.6c, d).

Closure

The uvula is approximated with 4–0 Vicryl sutures. The right-sided musculomucosal flap is inset using 4–0 Vicryl suture, and the left-sided mucosal flap is similarly inset (► Fig. 15.6e). Next, 4–0 PDS sutures are used to approximate the musculus uvuli and to overlap levator veli palatini muscles on each other. The 4–0 Vicryl sutures are used to inset the oral-lining z-flaps in a similar fashion (► Fig. 15.6f).

15.4.3 Postoperative Care

Cardiopulmonary monitoring is done routinely after palatoplasty. Many surgeons use elbow immobilizers in the postoperative period; however, data supporting their use are lacking. Postoperative diets vary among institutions. At our institution, clear liquids or breast milk via a cleft feeder is allowed immediately postoperatively. A full-liquid, blenderized diet is initiated on the first postoperative day, avoiding sharp utensils, straws, crunchy food, or other items that are potentially injurious to the repaired palate and is continued until postoperative day 10, and then, a soft diet is continued until postoperative week 6.

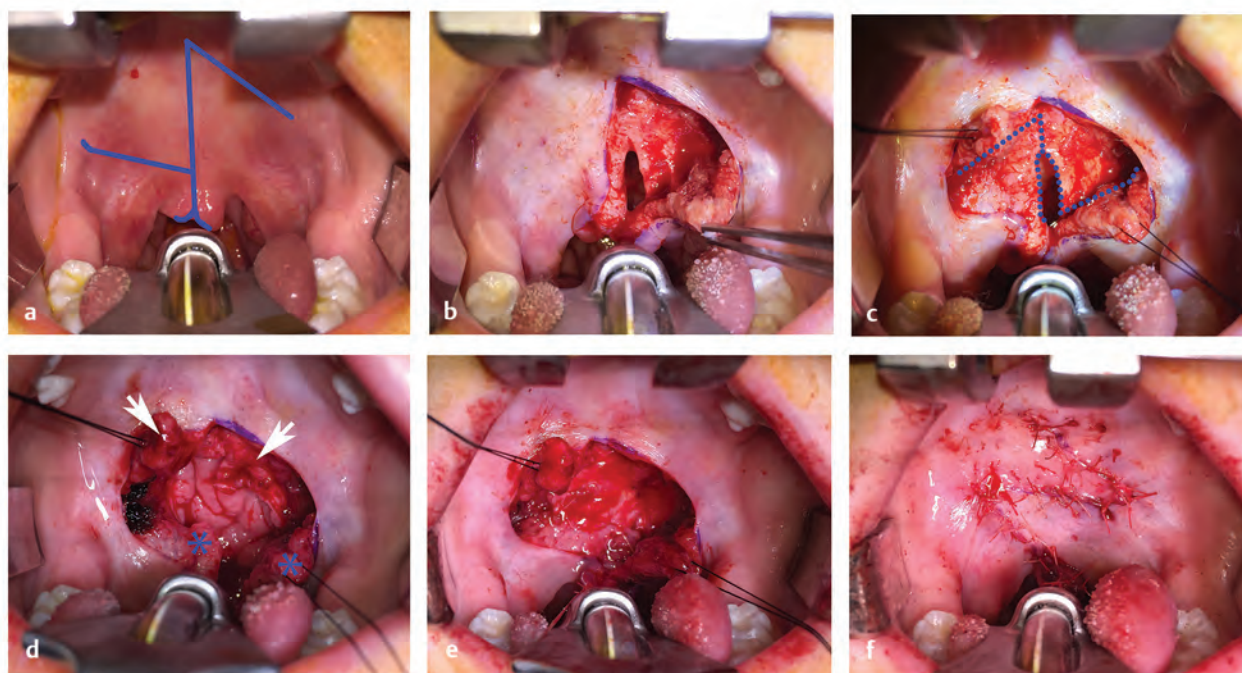


Fig. 15.6 Double-opposing Z-plasty (Furlow). (a) Oral-side rotation flaps are marked in blue in a patient with an overt submucous cleft palate. (b) The palate has been split, and the left posteriorly rotating musculomucosal flap is elevated. (c) The nasal-side rotation flaps have been marked, roughly in a mirror image of the oral flaps. (d) Musculomucosal flaps (*) and mucosal flaps (arrow) have been elevated. Note how the flaps orient themselves in position when properly designed. (e) The nasal layer has been closed with the musculomucosal flap posteriorly and the mucosal flap anteriorly. (f) The mirror-image oral flaps have been closed, similarly, with the musculomucosal flap rotated posteriorly and the mucosal flap rotated anteriorly.

The patient is discharged when he or she is medically stable and is taking enough liquid by mouth to stay adequately hydrated, typically on the first or second postoperative day. Follow-up varies by surgeon and cleft center. In the author's practice, patients are typically evaluated 7 to 10 days and 6 weeks postoperatively to assess the repair and help parents troubleshoot feeding issues. A full team evaluation with speech therapy and dentistry is performed at 20 to 24 months of age and annually thereafter.

15.5 Complications

Acute complications of the cleft palate repair are extremely rare but include dehiscence, bleeding, and airway-threatening edema. Some surgeons leave a stitch in the tongue for retraction in case of an airway emergency. Chronic complications include fistula formation (► Fig. 15.7), VPI (► Fig. 15.8), maxillary hypoplasia, and sleep apnea.

Incidence of palatal fistulae ranges from as low as 2 to 60% and is reported to be more common with higher Veau classification and repair at older age. Palatal fistulae occur at areas of tension in the repair, with the hard palate–soft palate junction being the most common site of unplanned fistulae. Minimizing trauma during dissection, eversion of the suture line under minimal tension, and good postoperative care are essential to preventing fistulae.

Velopharyngeal insufficiency results from inadequate closure of the velopharyngeal sphincter during speech, resulting in

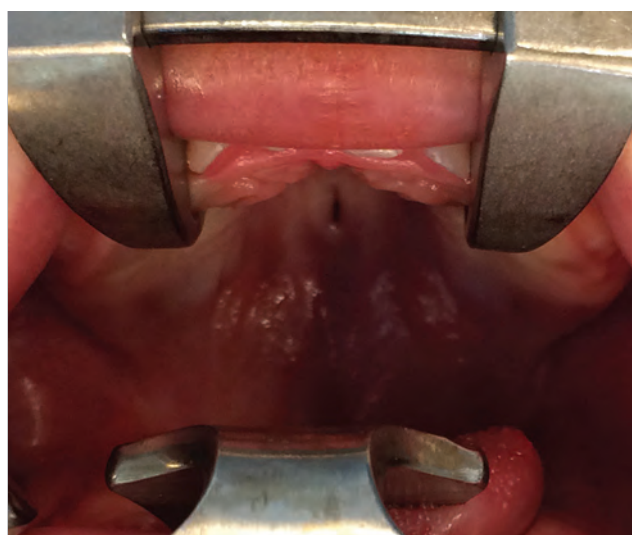


Fig. 15.7 Palatal fistula. Slit-like fistula in the central hard palate.

nasal air escape. This may result from a short palate or inadequate muscular repair but often has no clear cause. Incidence of VPI increases with greater Veau classification and increased age at the time of palatoplasty. The association between palatoplasty technique and incidence of VPI is unclear. With therapy, many children will be able to achieve adequate speech, but a small number of children require additional surgical

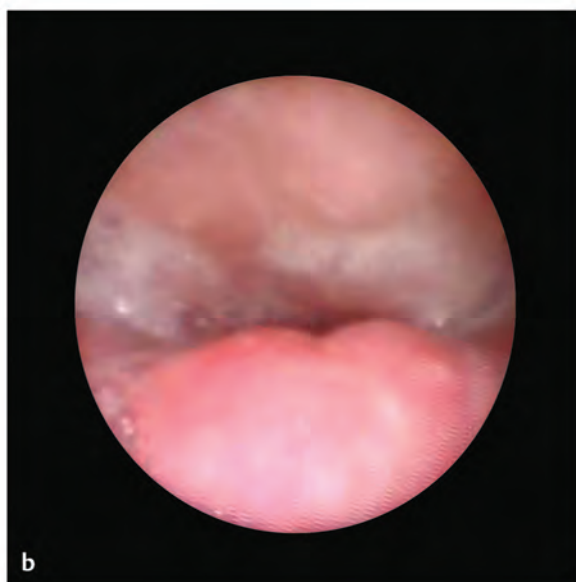


Fig. 15.8 Velopharyngeal insufficiency (VPI). **(a)** Nasal endoscopy demonstrates nasal air escape during production of plosives in a patient with mild VPI, as evidenced by air bubbles leaking at the left lateral aspect of the palate. **(b)** Nasal endoscopy demonstrates a large velopharyngeal gap in a patient with severe VPI.

procedures. These may include conversion to double-opposing Z-plasty, sphincter pharyngoplasty, pharyngeal flap, and augmentation of the posterior pharyngeal wall.

Children at the highest risk of developing sleep apnea are those with syndromic diagnoses and Robin sequence. Other

complications of cleft palate repair detected late in childhood include injury to tooth buds and maxillary hypoplasia. Maxillary growth deficiency, thought to be related to periosteal dissection, is most common in patients with bilateral cleft lip and palate, and maxillary hypoplasia is treated by Le Fort I advancement.

15.6 Conclusion

Successful palatoplasty hinges on separating the oro- and nasopharynx without fistula, achieving sufficient velar length, and creating a functional muscular sling. Successful comprehensive cleft care entails preoperative optimization of feeding, awareness and management of complications associated with palatoplasty, and multidisciplinary postoperative care to achieve the best functional outcome.

15.7 Key Points

- Cleft palate results from failure of fusion of facial prominences early in embryologic development. There are genetic and environmental influences. Isolated cleft palate is more likely than cleft lip and palate to be associated with an underlying syndrome.
- Cleft palate repair closes the oronasal fistula and establishes continuity of the soft palate musculature to optimize speech development.
- Secondary procedures may be required to address complications of surgery, such as poor speech and palatal fistulae.

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16 Robin's Sequence

Noopur Gangopadhyay and Kamlesh B. Patel

Summary

Robin's sequence is characterized by the triad of micrognathia, glossoptosis, and airway obstruction; in 50% of cases, a cleft palate is associated with the sequence. The cleft of the secondary palate is thought to arise from mechanical obstruction during the elevation of the lateral palatine processes from a vertical to a horizontal position. Conservative management of airway obstruction should be attempted first, with prone positioning, antireflux medications, and nasopharyngeal airway (NPA) as the mainstays of treatment. Surgical management is required in a small proportion of patients and should be preceded by airway evaluation by diagnostic endoscopy. Depending on the airway evaluation and relevant anatomy for each patient, tongue–lip adhesion, mandibular distraction, or tracheostomy may be required.

Keywords: Robin's sequence, micrognathia, retrognathia, glossoptosis, airway obstruction, mandibular distraction, tongue–lip adhesion

16.1 Introduction

Although initially described by St. Hilaire in 1822, the French stomatologist Pierre Robin receives credit for defining this disorder in 1923. Robin wrote more than a dozen articles describing a disorder consisting of micrognathia and glossoptosis, which resulted in airway obstruction and feeding difficulties. Unlike a syndrome in which multiple findings arise from a unifying pathogenesis, Robin's sequence arises from a chain of events, in which one anomaly leads to the next. The micro-retrognathia is thought to be due to an underlying genetic abnormality or intrauterine growth restriction related to deformation or an alteration in mandible position. This micrognathia then leads to posterior displacement of the tongue, resulting in airway obstruction and poor oral intake. Approximately half of the cases are associated with an isolated cleft palate, stemming from mechanical obstruction by the tongue during the elevation of the lateral palatine processes from a vertical to a horizontal position.

16.1.1 Epidemiology

The incidence of Robin's sequence is reported as 1 in 8,500 to 1 in 14,000 live births, though the estimates vary between 1 in 5,000 and 1 in 50,000 births. There does not appear to be a gender discrepancy. It usually presents in isolation and is referred to as a nonsyndromic Robin's sequence. A syndromic presentation occurs most commonly in association with Stickler's syndrome and 22q deletion syndrome, though several other syndromes share these findings. Mortality in the early half of the 20th century was reported as high as 50%, largely due to respiratory difficulties and aspiration. With improvements in diagnosis and treatment, mortality ranges from 1.8 to 30%, and is usually related to the severity of airway distress.

16.1.2 Etiology

The cause of Robin's sequence is unclear, and the majority of studies point to a multifactorial etiology. The micro-retrognathic mandible may be due to intrauterine growth restriction, as may occur with a multigravid pregnancy or oligohydramnios. External deformation results in constriction of mandibular growth forward. The tongue is then retropositioned in the mouth and inhibits elevation of the lateral palatine processes mechanically. Alternatively, delay of neuromuscular maturation can occur in the tongue and palate, leading to this sequence. Intrauterine exposure to ethanol and hydantoin has also been implicated.

16.1.3 Genetics

A genetic basis is heavily supported by the high incidence of twins with Robin's sequence and a higher frequency of cleft lip and palate in family members of patients with Robin's sequence. Deletions of 2q and 4p and duplications of 3p, 3q, 7q, 8q, 10p, 14q, 16p, and 22q are associated with cleft palate. Micrognathia is linked to deletions in 4p, 4q, 6q, and 11q and duplications of 10q and 18q. Benko has reported on an autosomal dominant locus of Robin's sequence on chromosome 17q24.3–25.1, which codes for regulatory elements of the *SOX9* gene, a key protein in chondrocyte differentiation. Rainger describes loss-of-function mutations in the *SATB2* gene, which also results in micrognathia and cleft palate in humans and in a mouse model.

Sonic hedgehog and Wnt signaling drive mandibular growth from the first pharyngeal arch during embryogenesis. Absence or downregulation of β -catenin reduces mandibular growth. Inactivation of leukocyte-antigen related (LAR) family receptor protein tyrosine phosphatase (*RPTP*) genes mimics the features of Robin's sequence in mouse model, resulting in aberrant jawbone and cartilage and decrease in cell proliferation in the mandible. Missense mutations in *DLX5* and *DLX6* in a canine model result in mandibular retrognathia and a cleft palate.

As early as 1978, Cohen recognized at least 18 syndromic diagnoses associated with Robin's sequence. This number has grown to more than 40 syndromes. In a systematic review of the literature, Izumi found that only 40% of cases were isolated Robin's sequence and that 60% of infants carried a syndromic diagnosis. Stickler's syndrome is the most commonly associated, with mutations in *COL2A1*, *COL9A*, *COL11A1*, or *COL11A2*, resulting in abnormal type II or type XI collagen; these two processes colocalize in 15 to 30% of patients. About 10% of patients with Robin's sequence are also affected by 22q deletion syndrome. This deletion in 22q11 leads to cleft palate, mandibular retropositioning, immune dysfunction, and cardiothoracic anomalies.

16.2 Diagnosis

The diagnosis of Robin's sequence still relies on the classical description from 1923—micrognathia, glossoptosis, and airway

obstruction. A cleft of the secondary palate is present in half of the cases. A multidisciplinary approach for the workup and care of these patients is required, with pediatric specialists in plastic surgery, otolaryngology, neonatology, gastroenterology, pulmonology, anesthesiology, speech pathology, and nursing all involved in diagnosis and treatment planning.

16.2.1 Presentation

Prenatal diagnosis is quite rare because micrognathia is often missed on a typical screening ultrasound. Micro-retrognathia is evident at birth and is the initial feature that prompts exploration for the diagnosis of Robin's sequence. Mandibular hypoplasia is present in the vertical and horizontal directions, and retrognathia—posterior displacement of the chin—is also notable. The degree of glossoptosis—posterior displacement of the tongue—is due to the structure and orientation of the mandible. The tongue, while of normal size, takes up relatively more space in a volumetrically smaller oral cavity. In addition, abnormal neuromuscular control of the tongue may lead to tongue prolapse. Obstruction of the posterior pharynx by the tongue results in occlusion of the airway during inspiration. This can lead to repeated episodes of oxygen desaturation, apnea, and cyanosis. Additional energy expenditure is required during breathing to overcome these forces, with use of accessory muscles; suprasternal retractions are common during these desaturation events. Furthermore, these episodes are not continuous given they may not manifest while the infant is awake but are common during sleep, while in the supine position, or during feeding. If untreated, this may lead to hypoxia, respiratory failure, and death.

Eating is particularly challenging for these infants, given the mechanics of breathing and feeding are in direct opposition. This can result in gastroesophageal reflux and aspiration. In addition, weight gain is poor because the energy required to breathe and feed often outpaces the caloric intake that these infants can acquire. Furthermore, in the population of infants with Robin's sequence who have a cleft palate, the inability to generate nutritive suckling in a cleft is compounded by micro-retrognathia and glossoptosis.

16.2.2 Adjunct Studies

Diagnostic fiberoptic endoscopy of the airway is critical to determine optimum therapy. This is useful to determine the level of airway obstruction, which may occur at the base of tongue, epiglottis, vocal cords, or in the subglottic structures. Laryngoscopy and bronchoscopy, if possible, should be performed under anesthesia by an otolaryngologist, to evaluate for laryngomalacia and tracheal stenosis as well as to assess for vocal cord mobility and dynamic airway changes. It is critical to perform these studies in different positions, given that prone positioning may resolve airway obstruction related to the base of tongue. In 1986, Sher described four types of airway obstruction in a cohort of 33 children, as noted on diagnostic endoscopy. In type 1 and type 2 patterns, the dorsal tongue is retropositioned, whereas type 3 and type 4 described lateral pharyngeal wall motion and pharyngeal stenosis, respectively, as the cause for airway obstruction. Sher's schema was then applied to infants with Robin's sequence and has some predictive and therapeutic value today. Sleep studies may also be

useful to evaluate for desaturation events during sleep, feeding, or position change, when the clinical diagnosis is not clear. It is not necessary to perform polysomnography routinely.

Cephalograms were used by Pruzansky to characterize mandibular shape and development in micrognathic patients; these cephalograms also give a sense of the posterior airway space. There is still controversy on whether the mandible stays persistently hypoplastic or undergoes a period of accelerated “catch-up” growth during the first few years of life. Airway obstruction often improves during this period of mandibular growth. A simple measurement of the distance between the maxilla and mandible in the horizontal occlusal plane can be used objectively to quantify the degree of mandibular hypoplasia. Both three-dimensional computed tomographic (CT) scans and three-dimensional surface photography are also useful in evaluating micrognathia and planning for surgical treatment.

16.2.3 Syndromic Robin's Sequence

Robin's sequence can occur in conjunction with one of a multitude of craniofacial anomalies in 60% of infants. ▶ Table 16.1 describes several of the familiar syndromes that are associated. Stickler's syndrome is the most commonly associated. This is usually an autosomal dominant disorder, notable for short mandibular height, myopia, and concomitant retinal detachment, hearing loss, and joint problems. An ophthalmologic examination is critical in infants presenting with myopia given that most infants are hyperopic, and this points to the diagnosis of Stickler's syndrome. 22q11.2 deletion syndrome is distinguished by a retrognathic mandible; cleft palate, including submucous clefts; impaired immune system function; hypotonia; and cardiothoracic malformations. Craniofacial microsomia and Treacher Collins syndrome, with their dysplasia of the midface and mandible, also occur with some frequency in patients with Robin's sequence. In several of these syndromes, associated midface hypoplasia compounds the degree of airway obstruction caused by glossoptosis. In these settings, a geneticist may be helpful to guide future family planning and to assist with expectations for long-term prognosis and development.

16.3 Nonoperative Management

Treatment of patients with Robin's sequence is usually dependent on the degree of airway obstruction. Conservative

Table 16.1 Common syndromic diagnoses associated with Robin's sequence

Syndromes		
22q deletion	Fragile X	TAR
Amniotic band disruption	Kabuki	Treacher Collins
Beckwith-Wiedemann	Marshall	Trisomy 13
Bruck	Moebius	Trisomy 18
CHARGE	Nager	Trisomy 21
Cornelia de Lange	Smith-Lemli-Opitz	VATER
Fetal alcohol	Stickler	Wolf-Hirschhorn

management is more effective in nonsyndromic than in syndromic patients with Robin's sequence. The two critical features are airway management and nutrition.

16.3.1 Airway Management

Prone positioning is the mainstay of therapy, relieving signs and symptoms in up to 70% of patients. This is quite effective when airway obstruction is localized to the base of tongue, given the prone position allows the tongue to fall forward. If feeding is not compromised, often no other treatment is indicated. This is successful in about half of infants with Robin's sequence. However, even these infants require rigorous observation because respiratory difficulty, feeding problems, and failure to thrive can develop over the course of the first few months of life.

More severe airway obstruction may require the placement of an NPA and/or supplemental oxygen. The NPA acts to bypass the site of upper airway obstruction given the distal end of the NPA sits beyond the base of tongue. Placement can be confirmed by endoscopy via the NPA. This method can be individualized for each infant, by modifying endotracheal tubes, chosen based on the infant's weight, as described by Chang. This allows for the introduction of supplemental oxygen as well. The care and cleaning of the NPA, as well as replacement, should be demonstrated to the parents to allow for home management. These adjuncts may be required for 2 to 4 months after discharge from the hospital.

Laryngeal mask airway or endotracheal intubation is the last resort to secure the airway in infants that do not respond to prone positioning or NPA. However, this is a temporizing measure indicating more severe obstruction and pushes the clinician toward surgical intervention for the patient.

16.3.2 Feeding

Even infants who do not exhibit respiratory difficulties when at rest may have airway distress while feeding. This is worsened if there is an associated cleft palate. In addition, syndromic patients may have underlying feeding difficulty or neuromuscular issues leading to difficulty in swallow coordination and poor oral intake. Increased metabolic rates require caloric supplementation by gavage feedings via a nasogastric tube. This can decrease the energy expenditure during feeding and facilitate appropriate weight gain. In severe cases, a gastrostomy tube may be required for feeding support. Evaluation by a speech pathologist and/or clinical dietician is critical for feeding support.

Gastroesophageal reflux in these infants can lead to airway inflammation, edema, and worsened airway obstruction. Empiric treatment is useful to improve feeding and breathing. In addition, sodium supplementation in infants has been effective in some infants to reverse failure to thrive. A weight gain of at least 20 to 30 grams per day is considered adequate.

16.4 Operative Management

16.4.1 Tongue–Lip Adhesion

Shukowsky was the first to describe tongue–lip adhesion (TLA) in the early 20th century, but the technique was popularized by

Douglas 40 years later. Tongue–lip adhesion serves to correct glossoptosis by pulling the base of the tongue forward and securing it to the lower lip. This mucosal attachment serves as a permanent tether, until the infant's growth results in a more stable airway. This must be done before the development of mandibular dentition. A second surgery is required to release the TLA when the airway is secure.

The GILLS scoring system, described by Rogers and Mulliken, is one measure that can predict the patients with Robin's sequence who will be good candidates for TLA. Five factors—gastroesophageal reflux disease (GERD), preoperative intubation, late presentation, low birth weight, and syndromic diagnosis—were determined to be significant in the success or failure of TLA. Again, airway obstruction and subsequent mandibular growth remain important factors for the outcomes in these patients, with TLA acting as a delaying type for the child to outgrow the glossoptosis.

This technique is appropriate after imaging has ruled out supraglottic and subglottic airway malformations. After nasotracheal intubation and supine positioning, a traction suture is placed through the anterior tongue. Mirror-image rectangular mucosal flaps are designed on the ventral tongue and lower lip lingual surface. The incisions are infiltrated with local anesthetic with epinephrine. The genioglossus muscle should be released subperiosteally from the lingual mandible via the ventral tongue incision. A permanent suture is passed around the mandible, through the lingual muscle, and buried across the alveolar ridge, as the tongue is pulled forward with the traction suture. The muscle and the mucosal flaps are then closed with absorbable sutures. The traction suture is secured to the lower lip and chin with adhesive strips; this can be pulled forward postoperatively in the setting of desaturations (► Fig. 16.1). The TLA is secured vertically to the external chin with two sutures and a button to relieve tension as the mucosal flaps heal. The traction stitch and button are removed 1 to 2 weeks postoperatively, and the TLA is taken down at 9 to 12 months of age.

This procedure is complicated by partial or complete dehiscence of the TLA, the need for reoperation, the need for a second surgery for TLA release, soft tissue infection, abscess, base of tongue scarring, and occasional conversion to tracheostomy. In addition, data on feeding in this population are mixed, with some cohorts experiencing worse outcomes due to altered tongue mobility and swallowing and other cohorts showing increased weight gain and a decreased need for nasogastric tube feeds.

Tongue–lip adhesion is appropriate for infants who fail prone positioning and whose airway obstruction is limited to the base of tongue. Success varies, with rates ranging from 36 to 100%. Rogers and Mulliken showed that patients with a GILLS score of 2 or less have a high success rate with TLA, whereas patients with a GILLS score of 3 or greater were five times more likely to fail TLA; using this stratification, they have shown a 90% success rate. However, patients with persistent symptoms and severe respiratory distress have a high rate of conversion to a second procedure such as mandibular distraction or tracheostomy in the first year of life.

16.4.2 Mandibular Distraction

Mandibular distraction osteogenesis (MDO) was described and popularized by McCarthy in 1989. It has become the mainstay

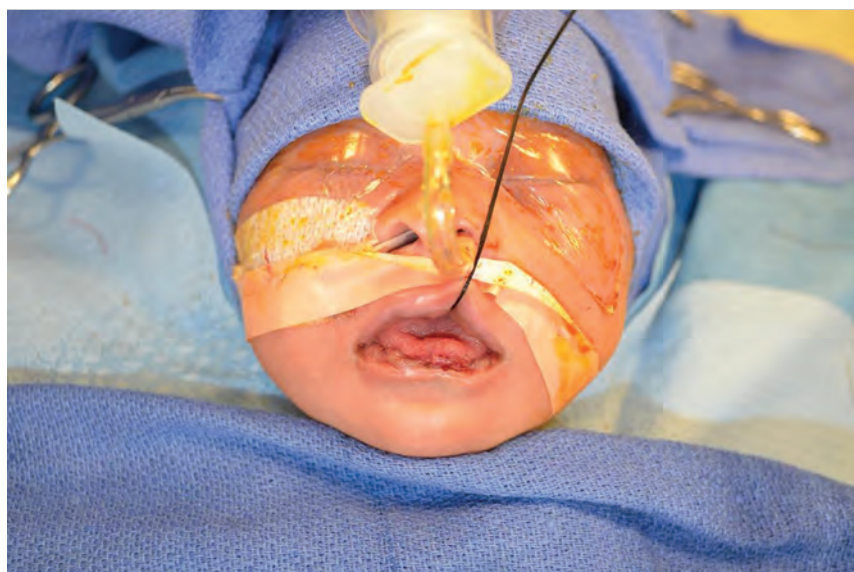


Fig. 16.1 Postoperative appearance of patient after TLA, with traction stitch and repair of mucosal flaps shown.

of surgical management for Robin's sequence, given that it addresses both airway obstruction and micro-retrognathia. The mandible is lengthened in an anteroposterior vector, pulling the base of tongue forward due to the lingual attachments to the mandible. Mandibular distraction osteogenesis requires bilateral mandibular osteotomies and placement of internal or external distraction hardware with adjustable percutaneous pins.

A preoperative CT scan can be advantageous to plan mandibular osteotomies, based on the shape of the mandible, potential tooth buds, and nerves. Denny describes the need for at least 17 mm of vertical height of the mandibular ramus to have adequate space for the osteotomy and device placement. There is significant variability in the orientation of mandibular osteotomies and modifications in technique with regard to distraction. Oblique osteotomies at the mandibular angle are quite common, whereas an inverted-L approach seems to minimize damage to tooth buds. The deficiency in Robin's sequence is in the horizontal dimension, so a horizontal or slightly oblique vector is preferred (though in cases associated with craniofacial microsomia, there are deficiencies in both dimensions).

The following technique, as described by Roy and Patel, illustrates a technique for MDO using internal distraction devices (Roy and Patel 2006). Fiberoptic laryngoscopy is performed before surgery and includes a jaw thrust to mimic MDO's end result. Bilaterally, the angle and inferior and posterior borders of the mandible are marked on the skin, and the mandible is approached 10 to 15 mm inferior to the border. Local anesthetic with epinephrine is injected in the dermis. The subcutaneous fat is swept superiorly and dissection proceeds deep to the superficial layer of the deep cervical fascia to avoid the marginal mandibular nerve. The pterygomasseteric sling is incised with electrocautery to expose the buccal cortex, and subperiosteal dissection frees the buccal and lingual surfaces of the mandible. An inverted-L osteotomy is performed over the vertical ramus of the mandible, at least 5 mm from the sigmoid notch. Internal microdistractors are secured with at least three 1-mm screws. Predrilling the holes is not required in infants. Activation of the mandible is performed intraoperatively to check for

completeness of osteotomies and hardware function. The distraction arm is brought through the skin posteriorly and inferior to the ear. The skin is closed in layers with absorbable suture.

The latency period can vary from 1 to 7 days, but most commonly, it is 2 to 3 days. Activation depends on patient's age and surgeon's preference. Distraction rates are 0.5 to 2 mm per day, spaced out in two to three intervals per day. The total amount of mandibular advancement varies from patient to patient, but the majority of craniofacial surgeons agree that overcorrection in the horizontal vector is critical. Respiratory symptoms and feeding are also evaluated as end points to completion of distraction. The consolidation phase is 4 to 6 weeks after the completion of distraction (► Fig. 16.2). Radiographic evaluation postoperatively is often obtained immediately after hardware placement, during the activation phase, and after hardware removal; delayed images are sometimes performed in follow-up to assess for relapse and mandibular union. A second procedure is required to remove the distraction hardware (► Fig. 16.3).

This procedure is complicated by pin site infections, unfavorable scarring, neuropraxia of the inferior alveolar nerve, marginal mandibular nerve palsy, damage to tooth buds, temporomandibular joint ankyloses, malocclusion, mandibular malunion or nonunion, hardware failure, and failure to resolve respiratory and feeding difficulties. Several series have described improvement in symptoms in up to 90% of patients, with avoidance of tracheostomy and successful decannulation of patients with tracheostomy in up to 70% of patients. Flores described superior outcomes when comparing MDO with TLA in nonsyndromic patients with Robin's sequence, regarding oxygen saturation and need for tracheostomy. In addition, there appears to be significant cost savings in the first 3 years of life for patients undergoing MDO as opposed to tracheostomy.

Mandibular distraction osteogenesis is appropriate for infants who have failed prone positioning and conservative measures and for those infants who would require a tracheostomy and/or gastrostomy in the neonatal period. Again, this is not appropriate for those infants who have subglottic airway anomalies.

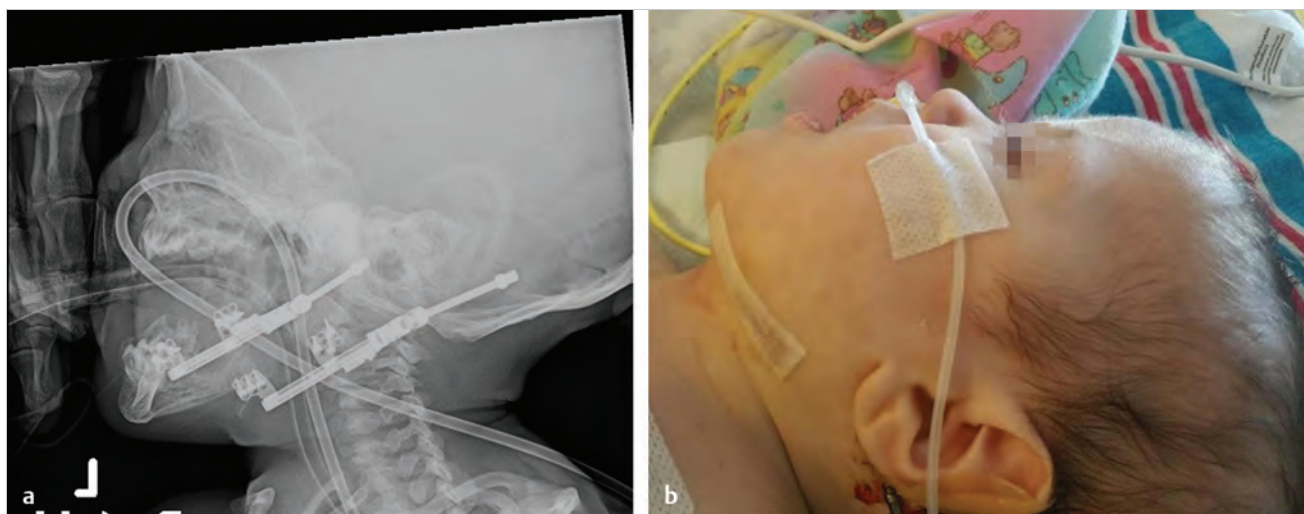


Fig. 16.2 Lateral radiograph showing device placement with distracted mandibular segments and clinical image of infant during the consolidation period.



Fig. 16.3 Preoperative image and postactivation image of patient after mandibular distraction.

16.4.3 Tracheostomy

Tracheostomy is often the last option for infants with Robin's sequence who fail conservative management and other surgical interventions. It remains the gold standard for definitive airway control and for those infants with subglottic stenosis,

tracheomalacia, or multilevel airway disease. The incidence of conversion to tracheostomy in infants with Robin's sequence is higher in patients with an associated syndromic diagnosis.

This procedure is complicated by bleeding, airway obstruction, tracheitis, pneumomediastinum, granulation tissue at the tracheal stoma, unfavorable scarring, unintentional

decannulation, tracheomalacia, subglottic stenosis, and frequent hospitalizations.

16.5 Complications

Hypoxia, desaturations, asphyxia, and aspiration are common without treatment in patients with Robin's sequence. In addition, feeding difficulties and recurrent respiratory infections are associated morbidities in Robin's sequence. Permanent brain damage and sudden death can result for apneic episodes. Mortality in patients with Robin's sequence ranges from 1.7 to 65%. Complications related to surgical technique are described earlier.

16.6 Conclusion

Care of the patient with Robin's sequence requires a multidisciplinary approach for diagnosis and treatment. Micrognathia, glossoptosis, and airway obstruction are hallmarks of this disease process, but the degree of airway obstruction and feeding difficulty drive treatment and prognosis. Treatment ranges from conservative measures such as prone positioning and nutritional support to surgical interventions, including TLA, MDO, and tracheostomy. The heterogeneity of presentations in nonsyndromic and syndromic cases requires an individualized approach to each infant with Robin's sequence.

16.7 Key Points

- Robin's sequence is a constellation of findings in which one malformation leads to the next.
- It is classically described as glossoptosis, micrognathia, and airway obstruction; a cleft palate is not necessary to make the diagnosis.
- Respiratory difficulties and feeding are important to address optimal management.
- Conservative treatment remains the mainstay of therapy.
- Surgical management includes TLA, MDO, and, as the last resort, tracheostomy.

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17 Secondary Cleft Lip and Nasal Reconstruction

Michael Alperovich and Roberto L. Flores

Summary

Secondary cleft lip and nasal deformities following primary cleft surgery represent some of the greatest reconstructive challenges and have inspired a multitude of creative technical solutions. Each secondary deformity is unique and must be evaluated independently with reconstructive plans grounded in principles rather than a formulaic approach. The number of described techniques is vast with limited evidence-based research to validate one method over another. However, techniques that are reliable in the experience of the authors will be presented to guide the plastic surgeon.

This review will discuss the important preoperative considerations in cleft lip and nose repair. Key physical exam findings will be presented. The philosophy of timing on secondary reconstruction will be explained including what the authors consider to be the major pitfalls in surgical approach.

Principles of secondary cleft lip reconstruction for unilateral and bilateral cleft deformities will be discussed followed by principles of secondary cleft nasal reconstruction of unilateral and bilateral cleft deformities. While an exhaustive review is impossible given the space limitations, this review will acquaint the reader with the major challenges and the available techniques in this field.

Keywords: cleft lip, cleft lip and palate, cleft, cleft rhinoplasty, cleft nasal revision, cleft nose revision, cleft lip revision

orthopaedics such as the Latham device are preferred by other centers. As described by Millard, dentofacial orthopaedics drives closer to completion the failed embryonic migration.

At the time of primary repair, exact intraoperative markings, precise incisions, and gentle handling of the tissue are all crucial for an optimal result. Cleft care delivered by high-volume centers with a multidisciplinary team approach is critical to maintaining excellent results. Minimum guidelines for cleft teams have been created jointly by the American Cleft Palate–Craniofacial Association and Cleft Palate Foundation.

Reports by McComb and Salyer demonstrated the safety and aesthetic advantage to primary cleft rhinoplasty. Long-term follow-up supports the overall safety of primary rhinoplasty on subsequent nasal growth. Although primary and revision cleft rhinoplasty are standard practice, great variability exists in approach. Plastic surgeons must remember that all primary repairs must consider the final on-table result in the context of long-term changes or the “fourth dimension” of time. Mulliken and LaBrie have characterized the specific anthropometric measurements that are altered in the fourth dimension. Cleft-side alar base drifts laterally (sn-al distance) and should be positioned more medially at cleft repair. Labial height (sn-cphi distance) lags on the cleft side and should be slightly greater on the cleft side at the time of repair. Finally, transverse labial width (cphi-ch) grows more on the cleft side and should be set shorter on the cleft side.

17.1 Influence of Primary Cleft Lip and Nasal Surgery

The first cleft operation is the best opportunity to achieve optimal results. Any revisionary surgery will be compromised by previously existing scars and violated tissue planes. When results are suboptimal at the initial surgery, revisionary surgeries become more challenging. Therefore, it is imperative to achieve the most aesthetic, functional, and lasting results at the first operation.

Our center has benefited from the implementation of nasoalveolar molding (NAM) into our presurgical treatment plan. NAM rotates the displaced alveolar segments into orthotopic position, lengthens the columella, increases nasal tip projection, folds the lower lateral cartilage toward its native form, medially transposes the lower lip and alar base, and stretches the intranasal lining. Multiple studies have documented the lasting benefit to nasal aesthetics following NAM and primary cleft lip and nasal reconstruction. Patients with unilateral cleft lip who undergo NAM demonstrate greater nasal symmetry through age 9. Patients with bilateral cleft lip who undergo NAM have a longer columella and near normal nasal morphology through 12.5 years of age.

Presurgical orthopaedics facilitates gingivoperiosteoplasty (GPP) and primary nasolabial fistula repair; however, NAM requires frequent follow-up and diligent care on the part of both the parents and the orthodontist. Alternative dentofacial

17.2 Timing of Secondary Cleft Lip and Nasal Surgery

In this chapter, we define rhinoplasty as primary when performed during the initial cleft lip repair, immature when performed after primary rhinoplasty but prior to facial maturity, and mature when performed at or after the time of facial maturity. Limited secondary cleft lip revision and immature rhinoplasty can be performed as early as age 5 or 6 prior to the child's entry into school to reduce social stigma during peer interactions. Early reconstruction is considered when patients have significant deformity or are suffering socially in peer interactions.

Because the nose grows until adolescence, rhinoplasty was often deferred until the later teen years. Ortiz-Monasterio and others have demonstrated excellent long-term follow-up even when rhinoplasty was performed prior to age 12. However, at this early age rhinoplasty precedes orthognathic surgery. Maxillary advancement alters the anthropometric dimensions of the nose, increases tip projection and nasolabial angle, and restores aesthetic balance to the face. As facial proportions are evolving and future rhinoplasty may still be required, the authors caution against comprehensive nasal surgery prior to facial maturity. Furthermore, patients prior to skeletal maturity may not have the cognitive maturity to verbalize their goals and concerns in immature rhinoplasty. At the time of facial maturity, patients become willing participants in their own care.

The great enemy of the refined soft-tissue repair is scar. Therefore, primary cleft rhinoplasty should achieve an aesthetic and lasting change using the least amount of dissection. The ideal is one rhinoplasty surgery during primary cleft lip repair and a second at the time of facial maturity. Active cleft surgeons are familiar with the scarred nasal tip resulting from excessive operations that resists the most elaborate technical maneuvers. The underoperated nose can be predictably improved although perfection is elusive. The overoperated, scarred nose can produce unfortunate, complex, and irreversible hurdles to an already challenging operation which, in many cases, can be avoided with proper planning, and an appreciation of the larger picture. Ultimately, the surgical philosophy of the authors is to effect the greatest change with the fewest number of surgeries and new scars. Diligence at the primary cleft lip and nose repair can reduce the number of surgeries required and avoid school-age interventions.

17.3 Secondary Correction of Unilateral Cleft Lip Deformities

Treatment of unilateral cleft lip deformities varies depending on the etiology of the deformity. Obtaining operative reports when the patient had the repair elsewhere may help with surgical planning, but in the authors' experience these reports add limited value compared to a good history and physical exam. Deficiencies can occur at each segment of the repair. Techniques will be characterized by vermilion deficiency, white roll misalignment, and a vertically short lip. When the deformity is significant, revising the entire cleft lip repair is the better intervention.

17.3.1 Vermilion Deficiency

Vermilion deficiency can result from insufficient attention during reconstructive planning of a primary lip repair, inadequate or incomplete repair of the orbicularis oris, or wound breakdown in the area of the vermilion. The two main categories of vermilion deficiency are loss of the vertical height of the vermilion and misalignment of the red line separating the dry from the wet vermilion. In Asians, African Americans, and Latino patients, discrepancies of the red line can be quite noticeable and therefore should be addressed at the time of lip repair. A modification of Noordhoff's triangular flap can restore the vertical height of the vermilion and reposition the red line.

During reconstruction, attention should be directed to careful repair of the orbicularis oris. Incomplete muscle repairs can result in loss of vertical height of the upper lip and vermilion. As scars in the vermilion tend to heal well, there should be a low threshold for excision of the affected vermilion and upper lip as part of the reconstructive plan.

For minor deficiencies isolated to the vermilion, fat grafting to the lip is a useful and minimally invasive technique that can be performed in the office. Overcorrection is preferred with an expectation that some fat will resorb, even in a well-vascularized bed such as the lip. Although the technique is reproducible, patients should be aware of the possible need for repeat surgery, particularly in the scarred vermilion.

Dermal grafting to the deficient vermilion also can restore volume. The most common criticism of dermal grafting is firmness and poor integration with the surrounding lip architecture. Local mucosal flaps have limited application outside mild deformities as deficiencies in this area are rarely isolated to the mucosa alone.

17.3.2 White Roll Misalignment

Misalignments of the white roll as small as 1 mm are noticeable at conversational distance. The step-off can be corrected with asymmetric Z-plasty or a diamond-shaped excision. Asymmetric Z-plasty can effectively align the two borders of the vermilion, but introduces a new oblique scar along nonanatomic landmarks of the upper lip. Patients with a predisposition for poor scarring are at risk for additional morbidity.

Diamond-shaped excision effectively lowers the white roll border to provide an upper lip height match in small discrepancies. As the incision line is hidden within the scar line of the previous lip repair, no new scars are formed. For this reason, the diamond excision technique is favored by the authors to correct minor discrepancies of the white roll.

White roll discrepancies of greater than 2 mm require re-creating the cleft deformity and repairing the cleft *de novo*. In these cases, the lip should be treated as a vertically short lip.

17.3.3 Vertically Short Lip

The vertically short lip can result from a planning error, technical error, or poor scar formation. Poor scar formation is related to patient biology and unlikely to be affected by revisionary surgery. Because poor scarring can haunt the cleft surgeon, careful physical examination should evaluate the presence of scar in other parts of the body or cleft repair. Poor upper lip scars share features of widening, placement lateral to the philtral line, and have wide areas of white roll disruption.

When there is a vertical discrepancy of greater than 2 mm between the cleft and noncleft philtral columns or when scars are wide and prominent, a cleft lip re-repair is advocated in the form of either a rotation-advancement or a straight-line repair.

Up to 25% of the upper lip can be excised without causing a disturbance in lip dynamics. Every effort should be made to completely excise all scarred areas of skin and muscle, restore vertical height of the lip, and locate the final scar along the philtral line.

Widened and prominent scars of the lip skin commonly overlie wide scars of the orbicularis oris. Incomplete muscular repair at the time of cleft repair may result in bulging of the affected upper lip with animation or vertical shortening. The surgeon should have a low threshold for resection of scar tissue in the orbicularis oris and complete reconstruction of the vertical height of the muscle.

17.4 Secondary Correction of Bilateral Cleft Lip Deformities

The secondary deformities affecting the unilateral cleft lip can occur in the bilateral cleft lip. However, the anatomic derangement is more complicated, as are the surgical techniques

required for correction. Due to space limitations, deformities related to alveolar clefts, displacement of the premaxilla, and oronasal fistulae will not be covered in this review. A malpositioned maxilla can have profound effects on nasal and upper lip form. The underlying bony foundation should be addressed prior to definitive lip or nose surgery.

Common bilateral cleft lip deformities include excessive scarring, a vertically short upper lip, and discontinuity of the white roll. Bilateral cleft lip repair requires the formation of two parallel, closely apposed incisions. Therefore, patients predisposed to excessive scarring will be at risk for suboptimal results even in the best of hands.

Some bilateral cleft lip repair techniques do not adequately reconstruct the entire orbicularis oris. In some cases, the muscle is not reconstructed at all. Absence of an intact oral sphincter interferes with creation of an appropriate prolabial width, adequate vertical height of the upper lip, and alar base position. The absence of orbicularis oris continuity is highlighted during puckering or whistling.

The midline tubercle of the vermillion should be assessed for presence and fullness. The height and width of the prolabium and the quality, location and area of the philtral scars are scrutinized and documented. Manchester reconstructions of the midline tubercle are antiquated, unaesthetic techniques that should be abandoned.

If enough unscarred philtrum exists, a bilateral cleft lip re-repair can adequately address the secondary deformities of the bilateral cleft upper lip. Provided that there is adequate tissue to create a columella-based philtral flap, surrounding scar affecting the upper lip skin, muscle, and mucosa is completely resected. If the lower lip is protrusive after orthognathic correction, the patient may be better served by an Abbe flap.

During bilateral cleft lip re-repair, vermillion turndown flaps are created from the lateral lip elements similar to primary lip repair. Care should be taken to reconstruct the entire length of the orbicularis oris at the midline. Wide dissection across the face of the maxilla may be necessary to relieve tension at the midline. Through this controlled reconstruction, vertical height of the upper lip and vermillion tubercle can be restored and unaesthetic scars replaced. Surgeons must anticipate additional widening of the prolabium with time. Philtral width is set narrower than desired to allow for stretching of skin with healing and activation of the orbicularis oris muscle. As will be discussed later, the forked flap advocated by Millard for primary bilateral cleft lip repair has no role in the modern techniques in cleft nasal reconstruction.

An Abbe flap can restore composite deficiencies in skin, muscle, vermillion, and mucosa to the midline upper lip in cases of excessively scarred or short philtrum (► Fig. 17.1). The Abbe flap serves to both augment the upper lip and reduce the lower lip volume, thereby restoring aesthetic balance to the mouth. The authors' preference is to perform the Abbe flap following definitive orthognathic surgery and secondary cleft rhinoplasty revision except in cases of severe distortion of the upper lip. The preference of the authors is to extend the Abbe flap onto the columella to avoid a depressed scar at the lip–columella junction. The fibrofatty tissue at the base of the prolabium is rolled onto itself superiorly to provide bulk and take the place of an absent anterior nasal spine.

The Abbe flap is designed narrower than needed for the same reason that the prolabial segment is created narrower at the initial bilateral cleft lip repair. Care is taken to prevent any vermillion misalignment of either the upper or lower lip during flap inset and donor site closure. Loosely tied wires can be placed between the maxillary and mandibular teeth if there is any

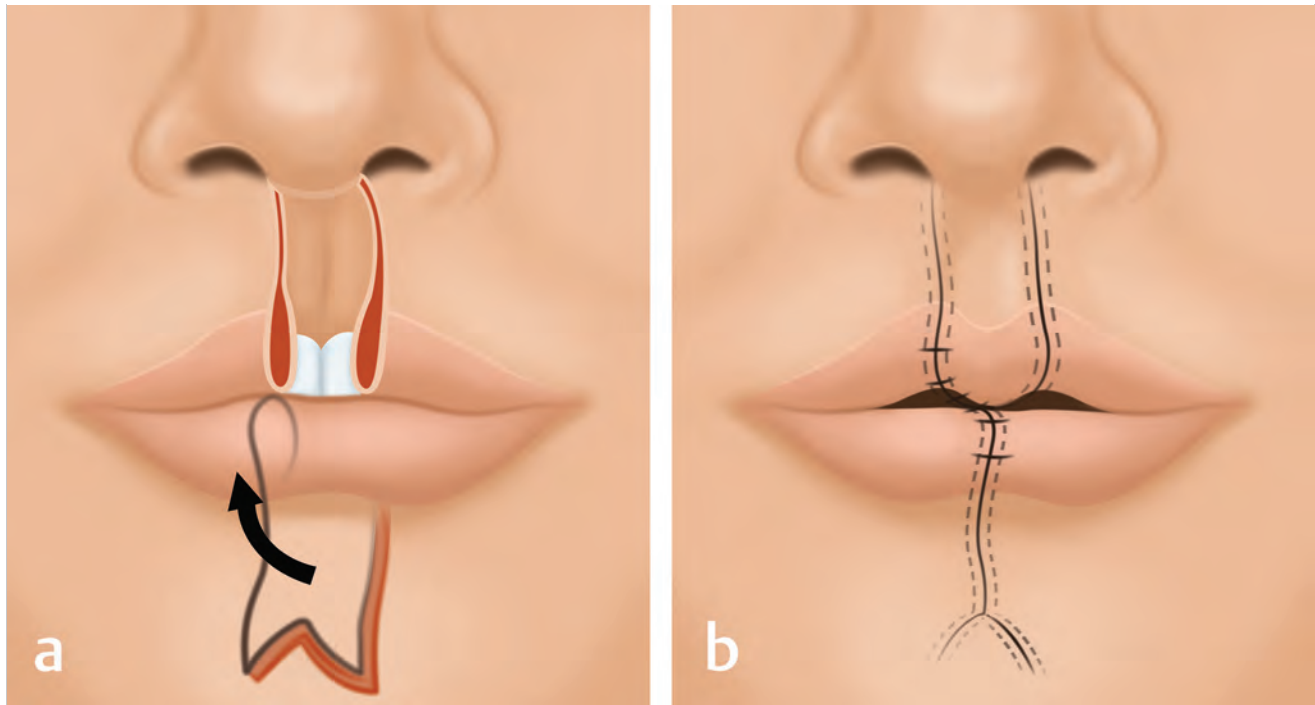


Fig. 17.1 Abbe flap for correcting deficient upper lip volume in a bilateral cleft lip deformity.

concern regarding patient compliance. However, given this procedure is typically performed in patients who are at least adolescents, there should be active involvement and investment on the part of the patient. Flap division is performed in an office setting one to two weeks later. The preference of the authors is to perform an Abbe flap in isolation without any other concomitant adjustments to preserve the integrity of the final result. However, other centers advocate combining the Abbe flap with a secondary rhinoplasty in the same stage.

As in unilateral deformities, fat and dermal grafting may be utilized for minor vermilion deficiencies that exist in isolation of other upper lip deformities. Similarly, V-Y advancement can be used to lengthen a deficient mucosa or vermilion although with limited efficacy.

17.5 Secondary Correction of Unilateral Cleft Nasal Deformities

Secondary reconstruction of unilateral cleft nasal deformity has evolved over the past several decades. Despite the significant variability in approach, most surgeons use the same set of key techniques. Mature cleft rhinoplasty usually is one of the last interventions for the cleft patient following maxillary advancement and alveolar bone grafting. A description of the unilateral cleft nasal deformity followed by commonly used techniques will be presented.

17.5.1 Unilateral Cleft Nasal Deformity Anatomy

The unilateral cleft nasal deformity is characterized by a deficient maxilla on the cleft side resulting in the cleft side alar base displaced posterolaterally and inferiorly. In a key historical study evaluating the anatomy of fetuses with cleft lip, the lower lateral cartilage of the cleft-sided nose was found to have distorted rather than deficient tissue. The cleft side lower lateral cartilage has a shortened medial crus and longer lateral crus. The angle between the medial and lateral crura on the cleft side is increased. The cleft-sided upper and lower lateral cartilages lack typical overlap. The nasal dome on the cleft side is retrodisplaced and less well-projected secondary to deficiencies in the maxilla. The nasal floor is also caudal on the cleft side and frequently absent. In addition, the lower lateral cartilage and alar rim create hooding. The ala is oriented posteriorly, laterally, and inferiorly on the cleft side relative to the noncleft side. The cleft-side nostril may buckle due to webbing of the vestibular lining. The shorter columella and caudal septum are deviated toward the noncleft side, while the bony septum is deviated to the cleft side. The nasal tip has a resulting asymmetry. Finally, the inferior turbinate on the cleft side often is hypertrophic.

17.5.2 Goals of Cleft Nasal Deformity Correction

The correction of the cleft nasal deformity in adults is generally less satisfactory than the cleft lip deformity. The pathological anatomy and reliable techniques are better characterized in the cleft lip compared to the nose. Furthermore, the lip repair is a

largely two-dimensional dilemma that follows an *ablative* approach to reconstruction; unacceptable and scarred lip is completely excised and fresh tissue is borrowed from laterally (or inferiorly in the bilateral cleft lip). The cleft nasal deformity is a three-dimensional dilemma where previous surgeries present *cumulative* obstacles in the form of scar and disrupted cartilage. There are virtually no options to replace scarred skin or mucosa from surrounding structures. Therefore, the most effective means of optimizing surgical results in the adult cleft rhinoplasty is to avoid the creation of excessive scar by overoperating on the nose during development.

The functional impact on the nasal airway is an often overlooked component of secondary rhinoplasty. Sixty percent of patients with cleft lip nasal deformity have some difficulty breathing. The obstruction is related to external nasal deformity, septal deviation, vomerine spurs, inferior turbinate hypertrophy, and maxillary hypoplasia. Secondary procedures such as pharyngeal flaps or sphincter pharyngoplasties also impact on airway resistance. Deviation of the bony septum can be an underappreciated contributor to nasal airway obstruction. The vomer is frequently deviated into the nasal airway on the cleft side and the perpendicular plate of the ethmoid can cause airway stenosis in the posterior recess of the nose. After submucous resection of the cartilaginous septum, additional resection of the bony septum may be required to restore airflow through the nose.

Multiple studies have developed assessment tools to evaluate outcomes following secondary cleft nasal deformity repair, but review of individual results remains largely subjective. The best study to evaluate impact on quality of life used analogue scales preoperatively and 1 year following secondary cleft rhinoplasty to measure psychological distress and generic quality of life. Interestingly, there was no correlation between subjective patient reported outcomes and surgeons' scores.

17.5.3 Evaluation

Surgical planning for immature and mature cleft rhinoplasty is dependent on an appropriately directed history and physical exam. History of all previous nasal surgery including previous rhinoplasty and septoplasty surgery is noted. Intranasal exam evaluates the presence of the cartilaginous and bony septum, septal deviation and dislocation, condition of the vomer and perpendicular plate, and the size of the inferior turbinate bilaterally. Airflow is assessed in both nasal airways with and without a Cottle maneuver. The skin and nostril rim are examined and palpated to assess the quality of the scar and the integrity of the lower lateral cartilages.

Visual inspection of the nose follows principles similar to traditional rhinoplasty. In the unilateral cleft rhinoplasty, dorsal deviation of the lower, middle, and upper third of the nose is closely scrutinized on frontal view. Lower lateral cartilage position and definition are noted as nasal tip cartilages are commonly widened due to inferior displacement and rotation of the cleft side lower lateral cartilage.

The nasal tip light reflex and the gullwing pattern of the lower border of the nose can expose asymmetries in the lower lateral cartilage position. The alar bases are compared for vertical and horizontal position. The lateral view of the nose is assessed on the right and left side as deficiency at the pyriform

aperture, tip projection, tip rotation, nasolabial angle, and columellar lobular angle can differ between the cleft and noncleft side.

In the basilar view, lower lateral cartilage position and deviation are inspected. Medial footplate position, columellar length, deviation of the columella and caudal septum, and nostril apex collapse are evaluated. The alar base position on the horizontal and anterior/posterior planes is noted, and the nasal floor is inspected for depression.

17.5.4 Techniques in Unilateral Cleft Nasal Deformity

The great majority of mature cleft rhinoplasty surgery is a secondary rhinoplasty involving extensive shaping of the nasal tip cartilages. As a result, an open technique is used in virtually all cases. Scar is the greatest impediment to successful manipulation of the nose in the mature cleft rhinoplasty. Cleft surgeons should *avoid overoperating on the nose*. Repeated “touch-up” surgery to the nose in the form of immature rhinoplasty will doom the patient (and surgeon) to a nose with fibrotic cartilage encased in an envelope of scar that permeates the nasal skin and mucosa and resists any meaningful manipulation. In order to combat the force of scar, particularly at the nasal tip, many surgeons advocate the use of costal cartilage grafts during rhinoplasty. In the authors’ experience, septal cartilage provides adequate rigidity in the majority of mature cleft rhinoplasty reconstructions.

Cutting grouped cleft rhinoplasty into bone, cartilage, and skin envelope techniques. Deviation of the upper third of the nose can be addressed with simple infracture and outfracture techniques. In certain cases, the perpendicular plate of the ethmoid is severely deviated, resulting in a tilt of the dorsal line of the bony vault. In these cases, the authors favor the monobloc osteotomy first described by Blair and Brown (► Fig. 17.2).

The technique requires a thorough resection of the deviated cartilaginous and bony septum. Low to low lateral osteotomies are employed followed by percutaneous osteotomy across the radix, leaving an intact bony bridge on the side opposite the

deviation. Firm, laterally directed pressure rotates the nasal unit as a single segment toward the midline. If the caudal septum is dislocated from the anterior nasal spine, it is separated, trimmed as needed, and secured to the midline with suture. The monobloc osteotomy can address the bony dorsum as well as deviations affecting the middle vault.

Deviation of the middle vault is addressed using spreader grafts. In cases of severe deviation, upper lateral cartilages are completely separated from the septum, and the entire middle vault is reconstructed to relocate the dorsal septum to the midline. Spreader grafts may be extended as a combined columellar strut to concurrently reinforce lower lateral cartilage position.

The emphasis of lower third nasal reconstruction is repositioning of the lower lateral cartilages. Columellar strut grafts are used often to reinforce the lower lateral cartilage framework. A floppy cleft-side lower lateral cartilage and scar within the skin and mucosa leave the lower third of the nose vulnerable to postoperative collapse. Although cartilage cap grafts commonly are used to camouflage nasal tip asymmetries, they will only exacerbate the deformity without rigid support from a columellar strut graft. Cartilage repositioning and reinforcement should be performed prior to any framework augmentation.

For mild asymmetries, the nasal tip may only require dissection of the lower lateral cartilages followed by suture reinforcement and a columellar strut graft. The tail of the cleft-side lateral crura commonly has fibrous attachments to the pyriform rim. These attachments should be completely excised prior to repositioning the lower lateral cartilage. For more severe asymmetries, composite rotation flaps from laterally or inferiorly can recruit cartilage toward the midline.

In cases where the alar base position is normal, lower lateral cartilage is recruited medially using a V-Y advancement flap first described by Potter. Intracartilaginous and intercartilaginous incisions are made on either side of the cleft-side lateral crura merging in a “V” at the lateral limit of the cartilage (► Fig. 17.3). The composite flap of cartilage and mucosa is advanced medially to restore tip projection followed by a V to Y closure of the defect.

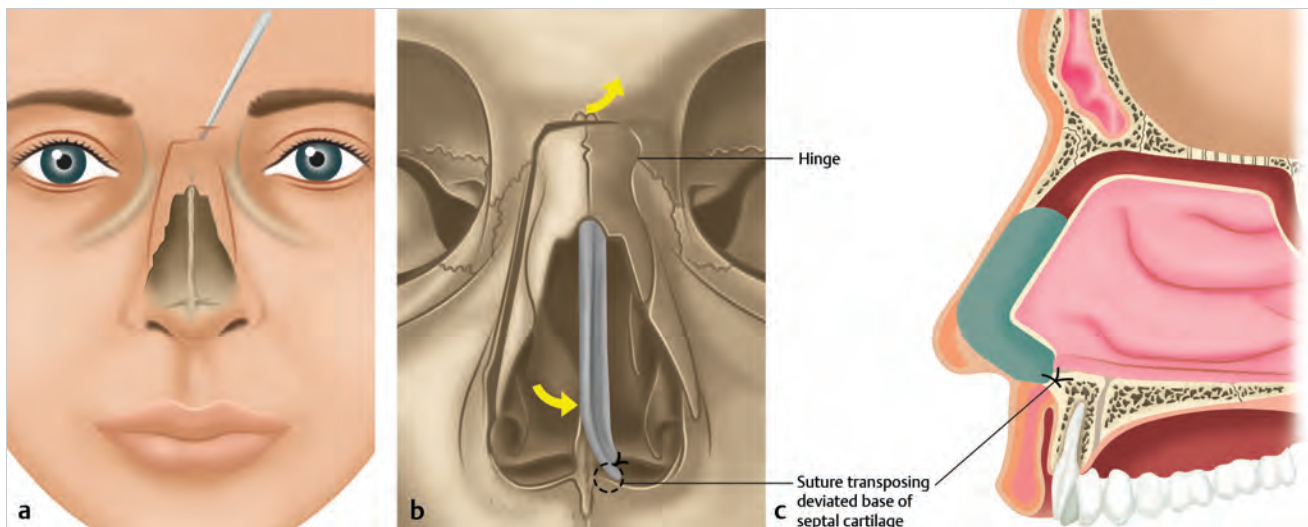


Fig. 17.2 Monobloc nasal osteotomy used to rotate the nasal bones and correct the upper third secondary cleft nasal deformity.

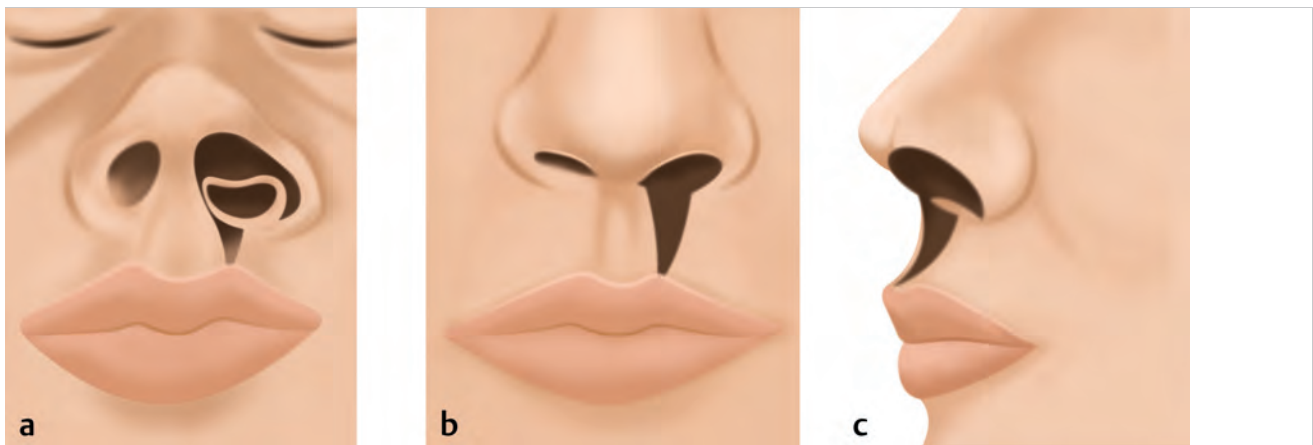


Fig. 17.3 The (a) Potter and (b) Dibbell procedures to correct the position of the cleft lower lateral cartilage in secondary unilateral cleft nasal deformity.

A group recently reported their experience with a modified V-Y chondromucosal composite flap similar to the Potter procedure, but which also included additional sutures and occasional use of a columellar strut graft. A shortcoming of the Potter technique is the cosmetic depression sometimes created by the transection of lateral ligamentous attachments.

In cases where the cleft-side alar base is in a lateral position and the nasal floor is depressed, the Dibbell procedure is used to recruit cartilage superiorly (► Fig. 17.3). Intercartilaginous and infracartilaginous incisions are extended medially as a hemimembranous incision with a skin incision anterior to the medial crura. These parallel incisions then extend medially just anterior and posterior to the cutaneous portions of the nostril floor and end laterally at the alar base.

This bipedicle composite flap is rotated superiorly, resulting in a medially transposed alar base, obliterated nostril floor depression, lengthened columella, and increased tip projection. This powerful technique should be combined with a columellar strut graft to reinforce the newly formed cartilaginous framework. The authors have found that excessive medial transposition of the alar base is possible with the Dibbell technique, and contralateral alar base resection may be required to restore nasal symmetry.

The typical skin envelope deformity in unilateral cleft nasal deformity includes nostril apex overhang, an absent soft triangle and a shortened columella. The Tajima reverse-U incision is a useful technique for tailoring and redraping the skin envelope after repositioning of the lower lateral cartilages (► Fig. 17.4). The incision is made inside the nose at the junction of the columella and membranous septum. The reverse-U incision is made around the superior border of the nostril rim to reflect the height and shape of the contralateral side. Wide subcutaneous undermining followed by suturing of the cleft lower lateral cartilage to the cleft and noncleft upper lateral cartilages as well as noncleft lower lateral cartilage is performed through a closed rhinoplasty incision. Finally, the reverse-U flap is thinned, reflected inferiorly, tucked within the nose, and inset to recreate the soft triangle. Although the Tajima technique uniquely increases the aperture of the nostril, precise markings are critical to success. An oversized flap will raid the nasal tip and rim, creating a contour deformity that is difficult to correct.

The authors prefer to use the Tajima technique during immature rhinoplasty and combine the Tajima with an open rhinoplasty during mature rhinoplasty. The open approach provides better visualization of the lower lateral cartilages and exposure for a columellar strut graft. The Tajima/open rhinoplasty can also be combined with a Potter or a Dibbell technique depending on the individual patient needs (► Fig. 17.4). Through the combination of different incisions and techniques, a customized approach to the lower third of the nose is used for each patient. This versatile approach is optimized through progressive experience with each of the listed procedures as a stand-alone operation and in combination with other techniques.

The micronostril is one of the most challenging deformities associated with the cleft nose. A result of excessive medialization of the alar base, lower lateral cartilage collapse, and excessive intranasal scarring, the micronostril lacks a reliable solution. Avoiding this deformity with a properly executed primary cleft rhinoplasty is the best treatment.

As lateral repositioning of the ala is difficult to perform without creating unaesthetic scars, the authors have relied more on camouflage techniques to correct the micronostril deformity. These techniques include a Tajima procedure to increase the aperture of the nostril on the affected side, alar base resection on the contralateral side, and repositioning of the columella toward the contralateral side.

17.6 Secondary Correction of Bilateral Cleft Nasal Deformities

The challenges of bilateral cleft nasal deformity correction lie in the restoration of tip projection, tip refinement, and increasing columellar length. Surgical correction is similarly limited by the force of scar upon the cartilaginous framework. Although the struggle of restoring three-dimensional symmetry to the nose is not typically present, the bilateral lack of tip projection, rotation, and definition require more extensive repositioning of the cartilage framework than the unilateral cleft nasal deformity. Given the decreased relative incidence of this phenotype, limited data exist regarding the techniques of repair. Following an explanation of the bilateral cleft nasal deformity, the principles in bilateral cleft nasal reconstruction will be discussed.

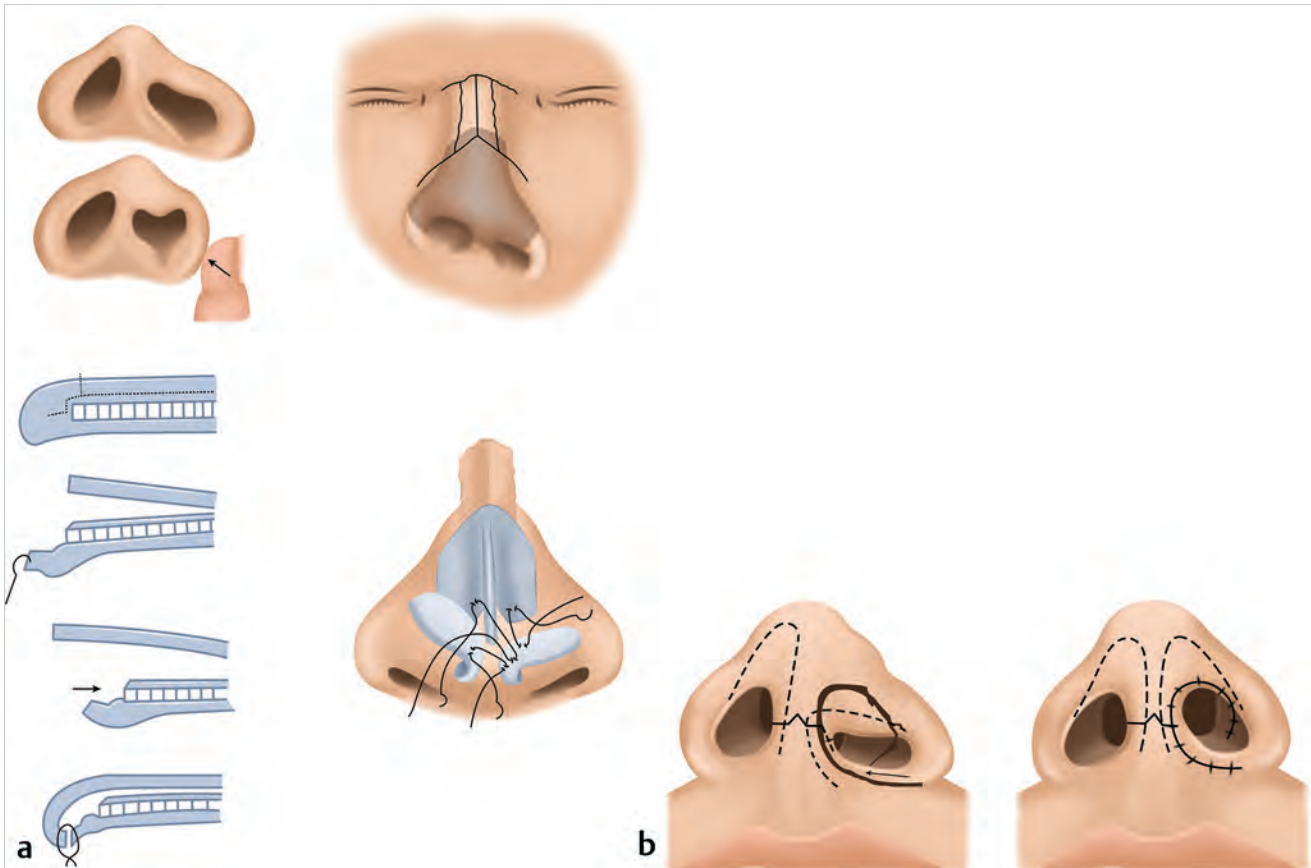


Fig. 17.4 The Tajima reverse-U (left) and cutting open rhinoplasty with combination Dibble/Tajima (right) procedures to address the cartilage and soft-tissue orientations in secondary unilateral cleft nasal deformity.

17.6.1 Bilateral Cleft Nasal Deformity Anatomy

The bilateral cleft nasal deformity is characterized by a widened inter-alar distance, shortened columella, lack of tip projection, and a broad and shallow tip. The prolabium can appear attached to the nasal tip. Similar to the cleft side in a unilateral cleft nasal deformity, the medial crura are shortened, while the lateral crura are longer with a hypoplastic maxilla bilaterally. The angle between medial and lateral crura is increased bilaterally.

The alae are displaced posteriorly, inferiorly, and laterally, and are flared in an S-shape. The vestibular lining of the alae has webbing bilaterally and the nasal floor may be notched. The caudal septum and anterior nasal spine are inferiorly displaced with fibrous attachments to the premaxilla. The septum is typically midline in the anterior aspect, but septal deviation can be present in the bilateral cleft lip patient.

17.6.2 Techniques in Bilateral Cleft Nasal Deformity

Bilateral cleft nasal deformity techniques seek to lengthen the columella, improve nasal tip projection, and narrow the alar bases. Cronin advocated columella lengthening by using skin from the nasal floor and ala. Forked flaps were popular for lengthening the columella, but resulted in a rectangular and

long columella, enlarged nostrils, and a sharp columellar–labial angle. These skin-based reconstruction techniques create excessive cutaneous scarring, do not restore the anatomic form or relationships to the cartilaginous framework, and, therefore, should be abandoned.

Modern techniques in bilateral cleft lip rhinoplasty emphasize alteration and reinforcement of the cartilaginous framework to restore nasal form. Nostril rim incisions, advocated by Mulliken, during primary rhinoplasty can be incorporated in immature rhinoplasty, particularly when nose surgery was not performed during primary lip repair. Tajima incisions may be used at this time although the authors caution the reader regarding the risks of overexcision of the nostril rim skin. At the time of mature cleft rhinoplasty, an open rhinoplasty approach is advocated with the use of Tajima incisions and Potter flaps as needed. As tip support is a challenge in bilateral clefts, columellar struts are obtained from septal or costal cartilage. Cutting favored an L-shaped septal cartilage graft as a spreader graft termed a “spreader-strut” to provide added projection. Onlay grafts to the tip can be applied in addition for volume.

17.7 Key Points

- The primary cleft repair is the best opportunity to achieve optimal results with any secondary surgery compromised by previously existing scars and violated tissue planes.

- Limited secondary cleft lip and nasal revision can be performed prior to the child's entry into school to reduce social stigma during peer interactions.
- In the presence of significant vertical lip height discrepancy or wide and prominent scarring, the cleft surgeon should not hesitate to perform a cleft lip re-repair.
- Rhinoplasty ideally occurs once at primary repair and again at facial maturity following any planned orthognathic surgery. The guiding principle is doing the most in the fewest number of surgeries.
- The majority of secondary cleft rhinoplasty surgery is performed using an open technique as it involves extensive shaping of the nasal tip cartilages.

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18 Velopharyngeal Dysfunction

Jenny T. Chen, Maia N. Braden, Joseph Lopez, Timothy W. King, Anand Kumar, and Howard Wang

Summary

Velopharyngeal dysfunction (VPD) describes the improper closure of the velopharyngeal valve leading to abnormal speech production including hypernasality and nasal air emission. VPD can be divided based on its underlying etiology into velopharyngeal incompetence (abnormal neurophysiology and movement), velopharyngeal insufficiency (VPI; abnormal anatomy), or velopharyngeal mislearning (articulation error). VPI frequently results from overt cleft palates, submucous cleft palates, or large palatal fistulas of the hard palate. Nonoperative management of VPI includes speech therapy and prosthetics such as palatal lifts and palatopharyngeal obturators. However, the primary treatment of VPI is surgical.

Keywords: velopharyngeal insufficiency, velopharyngeal dysfunction, velopharyngeal incompetence, velopharyngeal mislearning, sphincter pharyngoplasty, pharyngeal flap, Furlow double-opposing Z-palatoplasty, radical intravelar veloplasty

18.1 Introduction

The velopharynx is composed of the soft palate, or velum (from Latin which means veil), and pharynx. The velopharynx is made of five muscles: the levator veli palatine (LVP), the tensor veli palatine (TVP), the palatoglossus, the palatopharyngeus, and the musculus uvulae. The superior pharyngeal constrictors and the salpingopharyngeus contribute to pharyngeal wall movement. The pharyngeal plexus (9th and 10th cranial nerves (CN IX and CN X)) innervates all of these muscles, except for the TVP, which is innervated by CN V (► Table 18.1).

Closure of the velopharynx separates the oral and nasal cavities during both speech and swallowing. Opening of the

velopharynx is required for the consonants “m/n/ng.” Closure of the velopharynx is needed for the normal production of all other sounds in the English language. During closure of the velopharynx, the palate is pulled superiorly and posteriorly by the LVP and makes contact with the posterior pharyngeal wall at a condensation of the superior pharyngeal constrictors known as Passavant's ridge. Normal speech requires the rapid movement of the velopharynx in order to generate both oral and nasal sounds.

Velopharyngeal dysfunction (VPD) in individuals with cleft palate can result in audible stigmata, with hypernasal speech, nasal air emissions, and inappropriate compensatory articulation errors. The incidence of VPD after palatoplasty has been reported from 5 to 40%. The impact of VPD is profound, and has the potential to affect social function and self-image.

Although the most common cause of VPD is anatomic/structural abnormalities, any pathology that results in abnormal velopharyngeal function can cause VPD. Previous classification of VPD was filled with confusion. The etiology of VPD was categorized as anatomic, iatrogenic, or neurogenic. VPD was further divided into whether it resulted from structural, functional, mechanical, or dynamic palatal dysfunction. More recently, VPD is now classified into three major diagnostic types: velopharyngeal incompetence (abnormal neurophysiology), velopharyngeal insufficiency (VPI; abnormal anatomy), or velopharyngeal mislearning (articulation errors; ► Table 18.2).

18.1.1 Velopharyngeal Incompetence (Abnormal Neurophysiology)

This type of VPD is given to velopharyngeal pathology of unknown origin or pathology secondary to abnormal

Table 18.1 Muscles of the velopharynx

Muscle	Origin	Insertion	Innervation	Function
Levator veli palatini	Cranial base (petrous temporal bone)	Crosses the middle third of the soft palate fusing with the contralateral muscle to form the levator sling	CN IX–CN X	Superior and posterior elevation of the palate
Tensor veli palatine	Membranous wall of the eustachian tube	Pterygoid hamulus	CN V	Opens the eustachian tube
Palatoglossus	Tongue	Anterior velum	CN X	Depresses palate
Palatopharyngeus	Posterior and lateral pharyngeal walls	Velum via posterior tonsillar pillar	CN X	Lateral wall motion
Musculus uvulae	Posterior velum	Mucous membrane of the uvula	CN X	Upward movement and shortening of the uvula
Superior pharyngeal constrictors	Median raphe (posterior pharyngeal midline)	Medial pterygoid and the pterygomandibular raphe	CN X	Lateral wall motion
Salpingopharyngeus	Cartilaginous eustachian tube	Blends with the palatopharyngeus and constrictor fibers	CN X	Elevates palate

Abbreviations: CN IX, ninth cranial nerve; CN X, 10th cranial nerve.

Table 18.2 Types of velopharyngeal dysfunction

Type of velopharyngeal dysfunction	Definition	Examples
Velopharyngeal insufficiency	An anatomic or structural defect that prevents velopharyngeal valve closure	Cleft palate, submucous cleft, deep pharynx, adenoid abnormalities, hypertrophic tonsils
Velopharyngeal incompetency	A neurophysiological disorder in which poor movement of the velopharyngeal structures results in incomplete velopharyngeal valve closure	Neurogenic injury, hypotonia, apraxia, cerebral palsy, global developmental delay
Velopharyngeal mislearning	Articulation errors secondary to inappropriate opening of the velopharyngeal valve during attempts to produce oral speech phonemes	Lack of auditory feedback, compensatory speech production, developmental delay

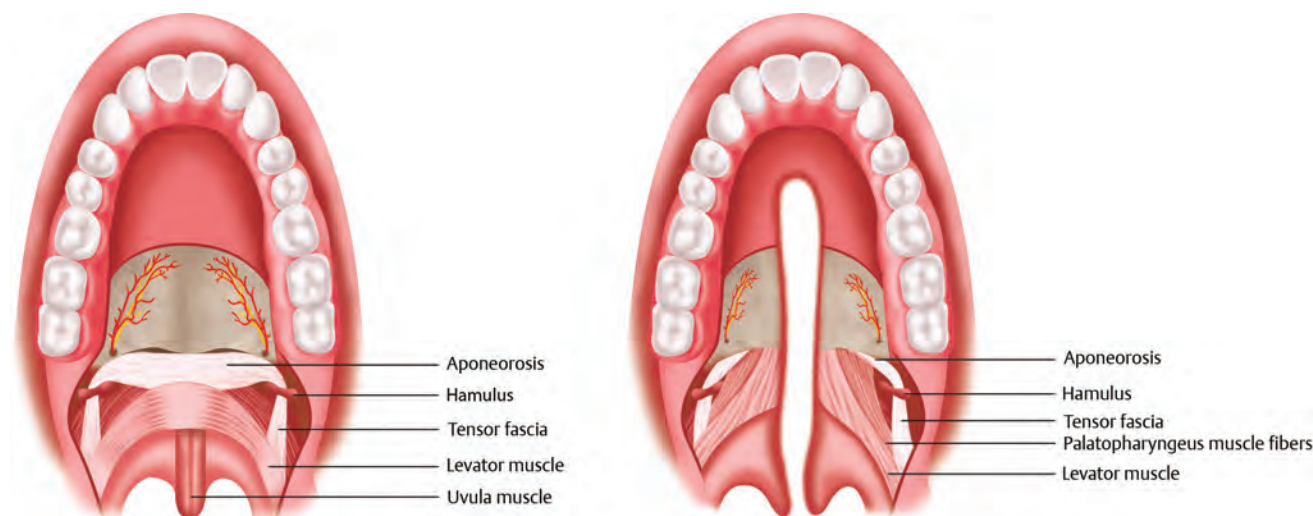


Fig. 18.1 Velopharyngeal anatomy. A figure demonstrating the significant differences in the normal velar anatomy compared to the cleft palate. Note the vertical position of both the levator and the tensor veli palatini muscles. The levator veli palatini muscle must be properly separated from the tensor muscle and transposed posteriorly into a more normal position to produce a more effective velar closing mechanism.

neurophysiology. It is important to note that in velopharyngeal incompetence, no underlying anatomical abnormality is present. The neurophysiological abnormalities associated with velopharyngeal incompetence can be caused by underlying intracranial processes, acute cerebrovascular injuries, palatal paralysis, or neurological injuries to the pharyngeal plexus. Velopharyngeal incompetence may also be associated with congenital neurologic conditions such as muscular dystrophy, hemifacial macrosomia, or cerebral palsy. The clinical manifestation of velopharyngeal incompetence is similar to other types of VPD but also presents with weakness, paralysis, or incomplete coordination of the velum and pharyngeal musculature, resulting in hypotonia, dysarthria, and loss of protective reflexes (i.e., gagging and swallowing).

In addition to neurophysiological abnormalities mentioned above, VPD can also present in patients with motor speech disorders, or apraxia. Since voluntary action of the velopharyngeal valve is mediated by coordinated signals from the motor cortex to the velopharyngeal musculature, it is not surprising that patients with apraxia can display abnormal coordination of the velopharynx during speech sound production.

18.1.2 Velopharyngeal Insufficiency (Anatomical Abnormalities)

The second major type of VPD is VPI. This categorization is given to VPD cases that are the result of structural or anatomical velopharyngeal pathology. VPI more commonly results from congenital abnormalities such as cleft palates, submucous clefts, or palatal fistulas (► Fig. 18.1). These congenital conditions are associated with gross tissue deficiency at the level of the velum and pharyngeal walls, and therefore limit velopharyngeal valve competence. Additionally, since velopharyngeal closure is largely a function of the ratio of the pharyngeal depth to palatal length, patients with disproportionately shorter palates or long pharynxes may also demonstrate incomplete closure of the velopharyngeal valve. Not only can VPI result from gross tissue deficiencies, or abnormal structural proportions, but it can also arise from congenital soft-tissue disturbances that produce mechanical dysfunction of the velopharyngeal valve. For example, hypertrophied tonsils can mechanically interfere with closure of the velopharyngeal valve and therefore cause VPI.

Lastly, VPI can stem from iatrogenic causes such as gross tissue deficiency secondary to tumor resection, trauma reconstruction, or adenoidectomy. Since removal of hyperplastic

adenoids is a common treatment for children with airway obstruction or chronic otitis media, adenoidectomy can commonly cause VPI. Conversely, procedures that increase the soft-tissue bulk of the velopharynx can also cause VPI. For instance, excessive soft tissue at the nasopharynx level as a result of a wide pharyngeal flap may limit apposition of the velum to the pharyngeal walls.

18.1.3 Velopharyngeal Mislearning

Patients with this type of pathology appear to have normal anatomical, neurogenic, and physiological velopharyngeal valve competency. However, these patients exhibit phoneme-specific nasal emissions; oral consonants are replaced by nasal airflow. Commonly, these patients display nasal emissions with only a select set of sounds such as CH, SH, Z, and S.

18.2 Diagnosis

Evaluation of VPD is necessary in order to identify the degree of dysfunction, the reason for the dysfunction, and to determine a course of appropriate treatment. Evaluation should be completed by a multidisciplinary team including a speech-language pathologist with training, knowledge and experience specific to the evaluation and treatment of VPD. Evaluation includes auditory-perceptual and instrumental assessments.

18.2.1 Perceptual Evaluation

Resonance

Resonance must be evaluated in speech. Resonance is a phenomenon that can only be perceived on vowels and voiced consonants. It is based on the amount of energy present in the oral cavity and/or nasal cavity. Hypernasality consists of too much energy in the nasal cavity on sounds that are not supposed to resonate in the nasal cavity. Hyponasality consists of too little acoustic energy resonating in the nasal cavity on sounds such as “m,” “n,” and “ng,” which are supposed to resonate in the nasal cavity. Cul-de-sac resonance, sometimes called “hot potato voice” is due to acoustic energy becoming trapped in the pharynx, sometimes due to large tonsils. Ideally, evaluators will be able to obtain a combination of structured and unstructured speech tasks. These should include a connected speech sample, sustained vowels, syllable strings of oral consonants and different vowels, syllable strings of nasal consonants and vowels, and words and sentences specifically useful in identifying a resonance disorder. Different rating scales of hypernasality and hyponasality are used, both clinically and in research. In a clinical setting, most speech-language pathologists use an equal-appearing interval scale, rating hypernasality on a scale of 0 to 4, or 0 to 5 of absent, mild, moderate, severe, and profound. Visual analog scales, direct magnitude evaluation, and a visual rate and sort method have all been used in the literature, and may be more sensitive and reliable than the equal-appearing interval method.

Audible Nasal Emissions

Audible nasal emissions are perceived when air escapes through the nose on a high-pressure consonant sound such as

“p” or “s.” They are due to incomplete closure of the velopharyngeal port during these high-pressure sounds, and the sound is thought to be due to turbulence either at the velopharyngeal port or in the nasal passages.

Articulation Errors

The speech pathology evaluation for VPD should always include an evaluation of articulation. This will identify speech sound errors that are developmental and unrelated to the VPD, but will also identify any obligatory or compensatory errors related to VPD. Individuals with VPD can have speech sound errors. These may be obligatory errors, such as nasalization of consonant sounds due to incomplete velopharyngeal closure. People may also develop compensatory errors, such as glottal stop substitutions, pharyngeal fricatives, and nasal fricatives, in an attempt to approximate normal speech sounds in the absence of a functioning velopharyngeal mechanism.

18.2.2 Instrumental Evaluation

Instrumental evaluation can be divided into indirect measures and direct measures. Indirect measures evaluate the speech output. Some examples of these include nasometry (Pentax), which measures the ratio of nasal acoustic energy to total acoustic output, and gives a percentage known as nasalance. This is a method of quantifying hypernasality or hyponasality. Pressure/flow measures are also used; these involve the placement of a mask and tube on the speaker's mouth and nose, to measure the airflow and air pressure from the mouth and nose during speech tasks. Other indirect measures used have included nasal accelerometer spectrography. Nasometry is the most widely used clinical measure.

18.2.3 Visualization

If perceptual and instrumental measures indicate that there is VPD, visualization of the velopharynx may be necessary in order to determine treatment. This is usually done by nasopharyngoscopy, but multiview fluoroscopy (MVF) is also widely used.

Nasopharyngoscopy

Nasopharyngoscopy is performed by the speech-language pathologist or physician. Topical lidocaine and oxymetazoline are often used for comfort. The procedure is not painful, and is generally very well tolerated. Very young children may be upset. Careful preparation and a calm demeanor are key to patient comfort and getting a good examination. The scope should be introduced to the child in a calm and age-appropriate manner, giving them plenty of time to become comfortable. In some clinics, the child is given a coloring book and other preparatory materials prior to the visit. Young children should be seated in their parents' lap, with someone available to steady their head as needed. Patient participation is extremely important, given we are evaluating the velopharyngeal mechanism during speech; some speech is necessary. The examiner should pass the scope through the nasal passage; passing through the middle meatus gives the best view. Once in place, and the VP port



Fig. 18.2 Instrumentation view. Nasal endoscopic view demonstrating the central gap in the soft palate associated with submucous cleft palate (SP), the velopharyngeal gap (VPG), and the posterior pharyngeal wall (PPW). (These images are provided courtesy of open access frontiers in Pediatrics, Front. Pediatr., August 11, 2014. <http://dx.doi.org/10.3389/fped.2014.00084>.)

can be fully visualized, the patient is asked to repeat a series of syllables, words, and sentences including oral and nasal sounds. Closure is observed during these speech sounds. This might reveal a submucous cleft palate, or incomplete closure for other reasons (► Fig. 18.2). Careful observation of closure pattern is necessary to determine the appropriate surgical intervention.

Multiview Fluoroscopy

MVF can also be used to evaluate velopharyngeal closure. This requires radiation exposure. If this is done, the child is positioned in the radiology suite and asked to repeat the speech stimuli in three positions: lateral, frontal, and base (Towne's) views. This technique gives a three-dimensional view of the velopharyngeal port, can provide actual measurements, and does not require insertion of a tube (camera). However, it does require a very cooperative patient, the study can take a long time as it requires each view to be taken independently, is more expensive than nasopharyngoscopy, and is most commonly only available in the hospital.

Functional Magnetic Resonance Imaging

Functional magnetic resonance imaging (fMRI) is emerging as a technology for visualizing velopharyngeal structure and function. With fMRI, imaging of the velopharynx is performed in four planes: sagittal, velopharyngeal axial (aligned perpendicular to the "knee" of the velum), axial, and coronal. Palate motion is shown using three-dimensional dynamic volumes created from the four views. Real-time visualization of velopharyngeal anatomy during its entire range of motion is possible. This technique, like MVF, also requires a very cooperative patient, is even more expensive than MVF, and is currently only available at a few specialized centers. It does, however, eliminate the exposure of the patient to ionizing radiation.

18.3 Nonoperative Management

18.3.1 Behavioral Speech Therapy

The speech-language-pathologist plays an important role in not only the assessment but also the management of patients with VPD. Behavioral speech therapy may be the primary treatment for patients with velopharyngeal incompetence or velopharyngeal mislearning. Speech therapy is also an important

adjunct to other treatment modalities for VPI and is often used in combination with surgical intervention.

18.3.2 Prosthetic Devices

Prosthetic devices are an alternative to surgical management. It may be used temporarily or serve as a permanent treatment option for patients who are not surgical candidates, such as those with sleep apnea or neuromuscular conditions. Several studies have shown similar speech outcomes with prosthetic devices as surgical management. Palatal lifts and palatopharyngeal obturators are the commonly used options. Each device is custom-made by the prosthodontist for the individual patient and is anchored to the maxillary dentition. Palatal lifts displace the velums superiorly to assist with velopharyngeal closure and is usually indicated in cases of neuromuscular dysfunction without anatomic deficit. In contrast, palatopharyngeal obturators have a posterior extension that occupies space in the velopharyngeal gap and may be used to augment closure of the velopharyngeal gap. The main advantages of prosthetic devices are avoidance of the complications associated with surgery and reduced cost. However, long-term use of prosthetics is complicated by noncompliance, need for frequent device adjustments, and emotional distress associated with wearing the prosthesis.

18.4 Operative Management

Surgical intervention is the primary treatment modality for patients with VPI due to an underlying anatomic abnormality such as those with a previous history of cleft palate repair. Commonly used surgical options for the treatment of VPI include radical intravelar veloplasty, Furlow double-opposing Z-platoplasty, posterior pharyngeal flap, and dynamic sphincter pharyngoplasty (► Table 18.3). Posterior pharyngeal wall augmentations have also been used to a lesser extent. Several clinical trials have attempted to compare outcomes of the different surgical approaches, but no significant differences have been identified. Each operation aims to improve velopharyngeal competence through a different mechanism. Thus, the choice of the operation should be selected based on the patient's pattern of velopharyngeal closure and the size of the remaining pharyngeal gap.

Velopharyngeal closure patterns can be classified as coronal, sagittal, circular, or bowtie. Pharyngeal flaps and sphincter pharyngoplasty both aim to decrease the size of the velopharyngeal port. Pharyngeal flaps bring tissue to the central portion of the velopharynx and thus are best utilized to correct central gaps such as with cases of sagittal or circular patterns of closure. Sphincter pharyngoplasty mobilizes tissue from the lateral aspects into the central portion and thus can correct lateral defects from coronal and bowtie closure patterns. Furlow palatoplasty lengthens the soft palate and tightens/repositions the levator sling. Radical intravelar veloplasty also tightens and repositions the levator sling. Both have been shown to be successful for small and medium central gaps less than 2 cm. Pharyngeal augmentation is also used at times to address small central defects less than 5 mm. Large "black hole" deformities are the most challenging to address, and strategies to do so include using static obturator, sphincter pharyngoplasty alone,

Table 18.3 Surgical treatment of velopharyngeal insufficiency

Operative techniques	Anatomic effects	Indications
Furlow double-opposing Z-palatoplasty	<ul style="list-style-type: none"> Lengthens the velum Reorients levator veli palatini into a transverse orientation 	<ul style="list-style-type: none"> Small to medium central gaps Primary and revision cleft palate repair
Radical intravelar veloplasty (IVV)	<ul style="list-style-type: none"> Lengthens the velum Reorients levator veli palatini into a transverse orientation 	<ul style="list-style-type: none"> Small to medium central gaps Primary and revision cleft palate repair
Posterior pharyngeal flap	<ul style="list-style-type: none"> Reduces the size of the airway with a static wall of mucosa from the velum to the posterior pharynx 	<ul style="list-style-type: none"> Large central gaps Revision cleft palate repair Requires adequate lateral wall motion
Sphincter pharyngoplasty	<ul style="list-style-type: none"> Decreases velopharyngeal port size by extension of the lateral and posterior pharyngeal wall 	<ul style="list-style-type: none"> Small to medium central gaps Poor lateral wall motion Requires adequate function of the levator veli palatini muscle

large posterior pharyngeal flap, or combining Furlow palatoplasty with sphincter pharyngoplasty. The varied treatment strategies for the large gap VPI is enigmatic and represents a current treatment challenge.

18.4.1 Radical Intravelar Veloplasty—Palatoplasty

Radical intravelar veloplasty is a well-described and useful treatment to treat the palate and optimize function from abnormal levator muscle position seen in submucous cleft palate and undertreated prior palate repair. This is incised in the midline from the junction of the hard/soft palate to the uvula with care to preserve the musculus uvulae muscle fibers (► Fig. 18.3a, b). The oral mucosa is elevated to the lateral palate shelves in order to visualize the entire levator veli palatini muscles, the tensor tympani muscles, and the tensor tympani aponeurosis bilaterally (► Fig. 18.3c, d). The tensor tympani aponeuroses is then transected and the abnormally positioned levator veli palatini muscles are then elevated from the nasal lining along with the fibers of the pharyngeal sphincters. The levator veli palatini muscles are then transposed to the midline and posteriorly at the level of the uvula and the central nonmuscular excess area is resected. The levator muscles, after they have been transposed posteriorly into a horizontal configuration, are overlapped and sutured together with the levator muscles under moderate tension (► Fig. 18.3e). The palate mucosa is then closed after hemostasis is achieved (► Fig. 18.3f).

18.4.2 Furlow Double-Opposing Z-Palatoplasty

Furlow double-opposing Z-palatoplasty is a versatile technique that leads to lengthening of the velum and repositioning of the fibers of the LVP into a transverse orientation, which enhances palatal mobility. The velum lengthening comes at the expense of decreased palatal width, making a tighter velopharyngeal port. The operation is performed by first making a midline incision along the soft palate and creating two Z-plasty flaps on the oral mucosa side of the velum. A posteriorly based oromuscular flap on one side and an oral mucosa-only flap on the opposite

side are elevated. During this dissection, care should be taken to avoid injury to the greater palatine arteries that supply the LVP muscles. After dissecting the oral flaps, a second set of nasal mucosa Z-plasty flaps are elevated. This time with one nasal mucosa-only flap on the side of the oromuscular flap and a nasomuscular flap on the side of the previously raised oral mucosa-only flap. The double Z-plasty flaps are then transposed and sutured in layers. A distinct advantage of the Furlow palatoplasty or radical intravelar veloplasty technique is that all other surgical options such as pharyngeal flap or pharyngoplasty remain available and may be used as an adjunct or salvage treatment for persistent VPI with this technique.

18.4.3 Posterior Pharyngeal Flap

The goal of the pharyngeal flap is to create a wall that connects the soft palate to the posterior pharynx, reducing the airflow through the velopharyngeal port. Nasal airway is maintained by the two lateral openings on either side of the flap, which implies that adequate lateral wall motion is critical to the success of the pharyngeal flap. First, a superiorly based myomucosal flap including the superior pharyngeal constrictor muscle is raised just superficial to the prevertebral fascia. Then, incisions are made at the nasal and the oral surface of the velum just posterior the junction of the hard and soft palate, and the two incisions are connected to create a pocket. The pharyngeal flap is then passed through the pocket in the soft palate and inset into the oral mucosa (► Fig. 18.4a). Primary closure of the donor site should be attempted. To maintain patency and control the size of the lateral ports, small endotracheal tubes or red rubber catheters are often inserted through each naris to maintain adequate airflow (► Fig. 18.4b). The soft palate triangular flaps are then inset to allow for complete closure and lining of the flap. Ideally, the pharyngeal flap should be inset at the level of the posterior nasal spine (PNS) parallel and in plane with the maxillary palatine plane defined by anterior nasal spine (ANS) to PNS to posterior pharyngeal wall (► Fig. 18.4c).

18.4.4 Sphincter Pharyngoplasty

Unlike posterior pharyngeal flap, sphincter pharyngoplasty creates an extension to the lateral and posterior pharyngeal wall.

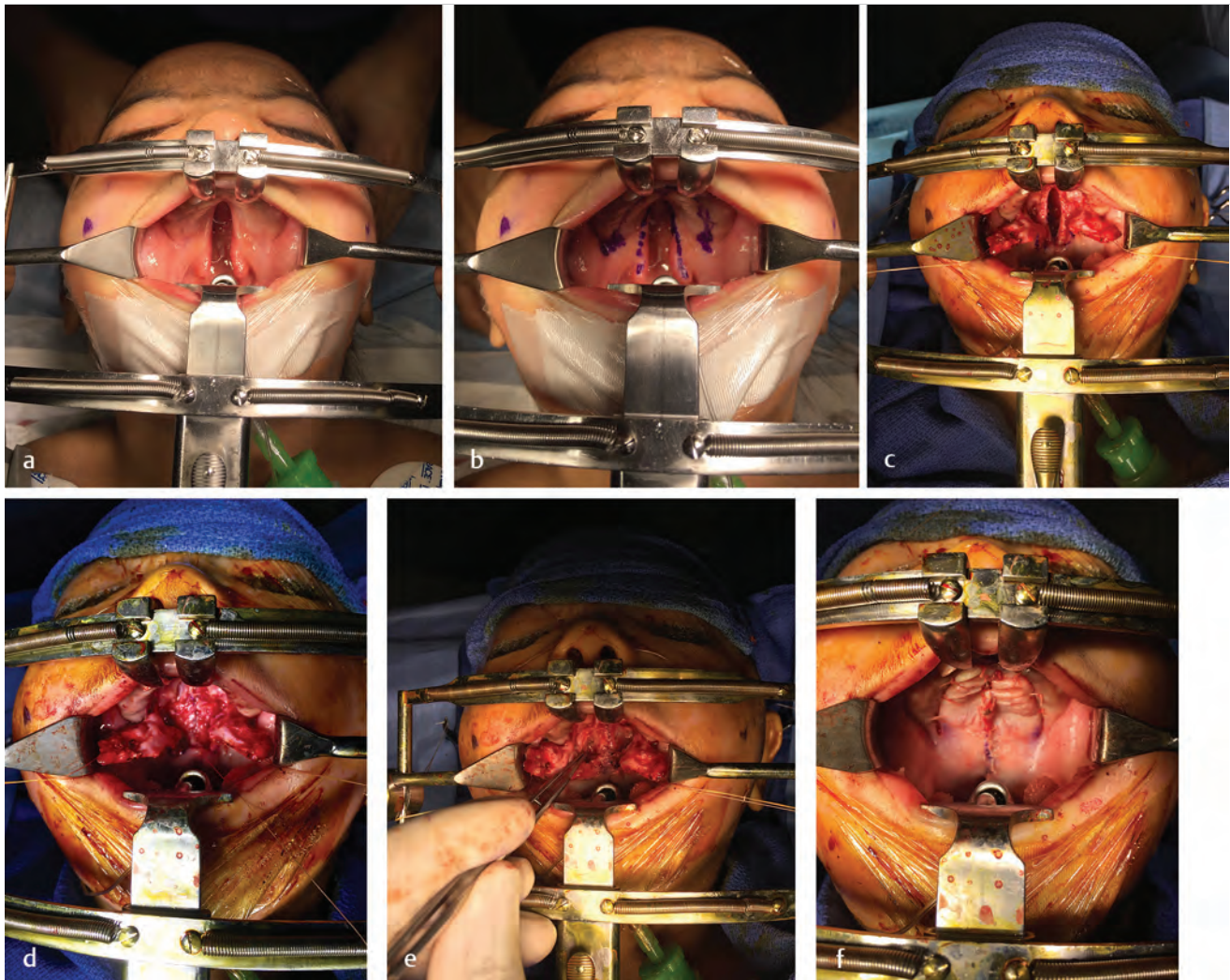


Fig. 18.3 Radical Intravelar veloplasty-palatoplasty. (a) Intraoperative view demonstrating a cleft of the hard and soft palate. (b) Standard markings for Bardach hard palate flaps and soft palate exposure. (c) Intraoperative view demonstrating elevation of the hard and soft palate flaps prior to closure. (d) Intraoperative view demonstrating closure of the nasal layer. Note the anterior position of the levator muscles after closure, in essence the creation of a submucous cleft palate. (e) Intraoperative view demonstrating proper repositioning of the levator muscles into a horizontal position in a proper radical intravelar veloplasty (IVV) procedure. The tip of the Gerald forceps is located at the junction of the lateral and vertically placed tensor veli palatini and the horizontally placed levator veli palatini muscles. (f) Intraoperative view demonstrating final appearance of the repaired cleft palate after closure of the hard palate and oral layer of the soft palate.

Thus, adequate function of the LVP muscle to close the central port during speech production is important for the success of this operation. The operation is performed by elevating bilateral superiorly based myomucosal flaps of the palatopharyngeus muscle. The flaps are then pivoted medially as high as possible on the posterior pharynx. Achieving high inset leads to improvements in speech outcomes but may be impeded by the presence of prominent adenoid pad. In such cases, partial or complete resection of the adenoid pad may be considered prior to sphincter pharyngoplasty. A transverse incision is made on the posterior pharynx where the two flaps are then sutured together either end to end or overlapped and inset onto the posterior pharyngeal wall. Primary closure of the donor site should be performed to decrease risk for cicatricial contracture, which can lead to inferior displacement of the myomucosal flaps. A suggested advantage of sphincter pharyngoplasty is

that the sphincter formed by bilateral palatopharyngeus muscles may have some dynamic function, allowing it to make adjustments during speech to avoid hypernasality and hyponasality that may result from imperfectly sized flaps. However, these claims have not been clearly proven.

18.4.5 Posterior Pharyngeal Wall Augmentation

Posterior pharyngeal wall augmentation through the injection of a variety of alloplastic or autologous material have been described. The goal of these techniques is to bring the pharyngeal wall closer to the velum to aid closure during speech. Most techniques have shown inconsistent success and carry risks for numerous complications including infection, extrusion, migration, and even embolism. With the increasing popularity of fat

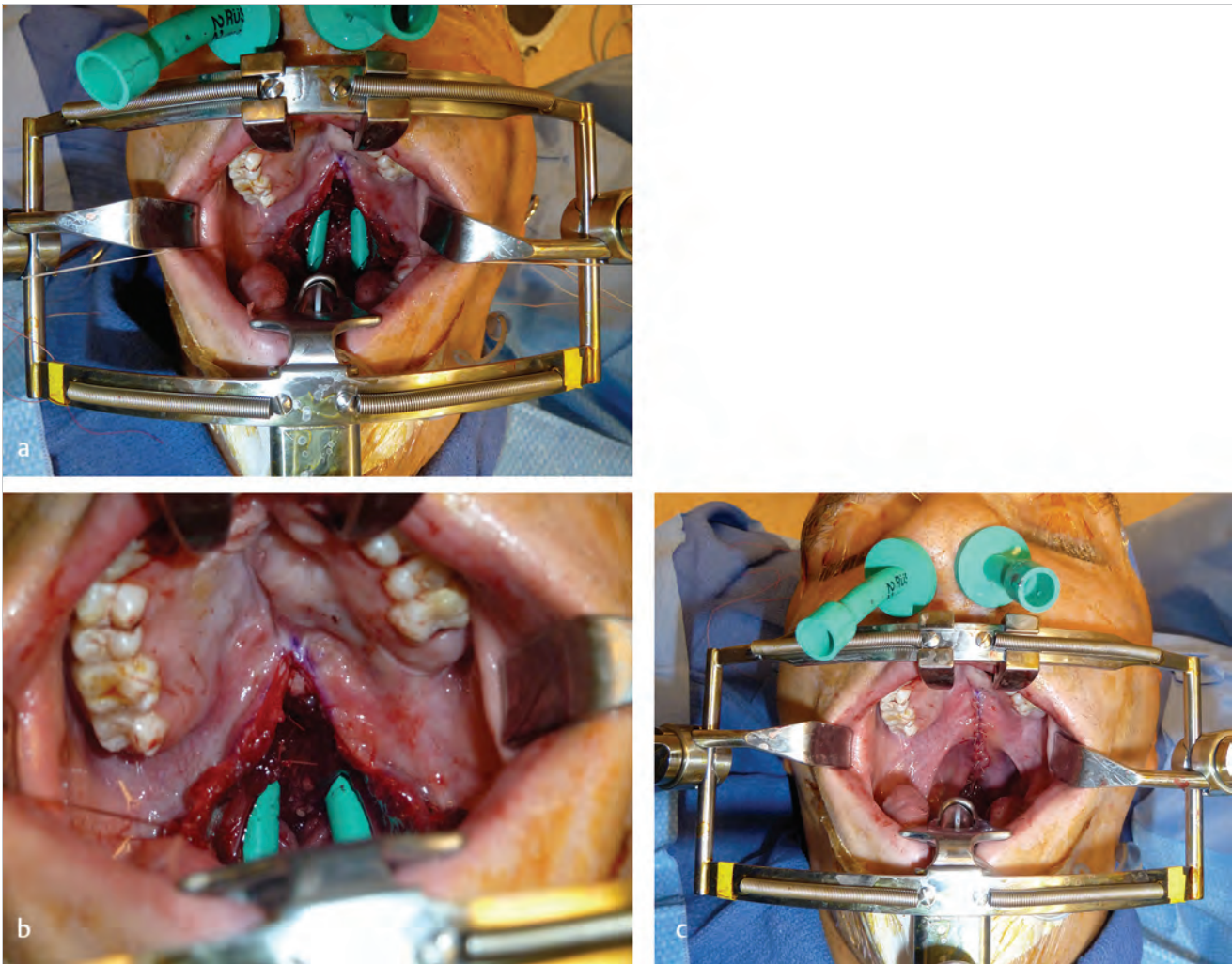


Fig. 18.4 Posterior pharyngeal flap. (a) Intraoperative view of a pharyngeal flap procedure after the flap has been elevated and sutured anteriorly to the hard/soft palate junction. The green nasopharyngeal (NP) airway tubes clearly demonstrate the lateral ports that have been created. (b) Intraoperative apical and magnified view of the lateral ports created after the posterior pharyngeal flap has been placed. We use various sized NP airway tubes to assist with port size creation and visualization. (c) Intraoperative view demonstrating closure of the soft palate after posterior pharyngeal flap placement. Note the normal appearance of the soft palate. We emphasize high placement of the pharyngeal flap at the level of the maxillary occlusal plane (ANS-PNS) to avoid complications of stenosis, inferior displacement and sleep apnea.

grafting, injecting autologous fat for pharyngeal wall augmentation has been reported by several authors in recent years. Its efficacy remains unproven, but it may have promise as an adjunct to palatal lengthening in severe cases of VPI.

18.4.6 Treatment Algorithm

One treatment algorithm that we use is based on closure pattern and gap size. If there is an unrepaired submucous cleft palate or a lack of posterior translocation of the levator veli/pharyngeal constrictor complex or palatal to pharynx gap at rest is less than 10 mm, a revision cleft palate procedure is performed typically a revision radical intravelar veloplasty or double-opposing Z-palatoplasty based on the surgeon's preference and training bias. In cases with a similar closure pattern but the gap is greater than 10 mm, a superiorly based pharyngeal flap would be preferred. If VPI is due to poor lateral wall motion with adequate levator function and gap is less than 10 mm, then

dynamic sphincter pharyngoplasty is chosen to address the lateral deficiencies. A combination of revision cleft palate repair and with dynamic sphincter pharyngoplasty is performed for gaps greater than 10 mm in the context of poor lateral wall motion and lack of proper levator muscle position. If there is a circular closure pattern but persistent gap of less than 10 mm, a double opposing Z-palatoplasty is then performed. With gaps greater than 10 mm, a pharyngeal flap is chosen.

18.5 Complications

VPD, if untreated, leads to hypernasality, nasal air emission, and poor articulation. While the use of prosthesis will avoid the complications associated with an operative intervention, it is not without any complications. For example, palatal prosthetic use leads to poor dental hygiene and may accelerate development of dental caries.

Surgical management of VPI with either radical intravelar veloplasty or Furlow double-opposing Z-palatoplasty is associated with inherent complications of any palatal surgery, which include flap loss and development of oronasal fistulae. Furthermore, it may result in insufficient gain in velum length and residual hypernasality. Pharyngeal flaps are static and thus its efficacy depends on the precise design of the size of the flap. If it is too narrow, there may be persistent nasal airway emission and hypernasality, whereas a flap that is too wide may obstruct the passage and lead to hyponasality or obstructive sleep apnea (OSA). Medial displacement of internal carotid artery can occur in patients with velocardiofacial syndrome; thus, it is important to obtain preoperative vascular imaging if posterior pharyngeal flap is planned in this patient population.

OSA may be a late complication of any surgery for VPI. Pharyngeal flap in particular is associated with a higher incidence of OSA; thus, signs and symptoms of OSA including snoring, daytime sleepiness, hypertension, and difficulty sleeping should be noted. Intraoperative corticosteroid administration should be considered to reduce postoperative edema. OSA that persists past the postoperative period may require continuous positive airway pressure or surgical treatment including division of the pharyngeal flap or tailored orthognathic surgery. Formal polysomnography may be required.

18.6 Conclusion

VPD can result from a broad range of etiologies and can be divided into velopharyngeal incompetence, VPI, or velopharyngeal mislearning. VPI results from anatomic abnormalities and is common after cleft palate repair. Evaluation should include perceptual speech assessment and instrumental assessments of the pharyngeal closure mechanics. While no one operation have demonstrated clear superiority, the choice of operation should depend on the patient's velopharyngeal closure pattern. When properly performed, the commonly performed operations for correction of VPI all have shown significant improvement in speech outcomes with a low risk of complications.

18.7 Key Points

- The etiology of VPD can be categorized into abnormal anatomy (VPI), abnormal neurophysiology (velopharyngeal incompetence), or articulation error (velopharyngeal mislearning).
- The diagnosis and management of VPD requires a multidisciplinary approach including speech-language pathologist, pediatric otolaryngologist, and craniofacial surgeon.
- Surgical treatment of VPI include radical intravelar veloplasty, Furlow's double-opposing Z-palatoplasty, posterior pharyngeal flap, sphincter pharyngoplasty, and pharyngeal augmentation.
- Selection of treatment option should be based on the closure pattern and the size of the remaining gap.

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Part III

Soft-Tissue Deformities of the Head and Neck

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19 Soft-Tissue Injuries

Arin K. Greene

Summary

Lacerations are common in the pediatric population. Iatrogenic injury includes intravenous extravasation during hospitalization. Management of soft tissue injuries depends on the etiology of the problem.

Keywords: dog bite, laceration, intravenous, extravasation, pediatric, soft tissue, trauma

19.1 Introduction

Soft-tissue injuries are one of the most common conditions treated by plastic surgeons. Parents, pediatricians, and emergency room physicians are particularly sensitive about scarring on a child. Consequently, plastic surgeons are frequently consulted to manage pediatric facial lacerations. Because wound management is the core of our specialty, plastic surgeons also are commonly asked to manage difficult and/or iatrogenic soft-tissue deficits throughout the body (e.g., extravasations, pressure ulcers, infections, etc.). Management of pediatric soft-tissue injuries is based on the principles of wound healing. This chapter will focus on two of the most common consultations for soft-tissue injuries: facial lacerations and intravenous extravasation injuries.

19.2 Diagnosis

Soft-tissue injuries are evaluated by history and physical examination. Identification of the etiology of the wound is important because it will affect management. For example, if the facial wound was caused by a dog bite, the individual is at greater risk for infection compared to being lacerated by a clean piece of glass (► Fig. 19.1). The depth of the injury must be ascertained. Blood often camouflages the area and should be removed gently to identify the severity of the injury. A partial thickness wound and/or abrasion may not require intervention. In contrast, a full-thickness injury might necessitate laceration repair. Patients at risk for a foreign body should have the area visually inspected. Plain radiography may be indicated to rule out material in the wound. Soft-tissue injuries in children are managed differently than in adults. Patients usually require sedation and are more likely to have absorbable sutures because they less able to tolerate suture removal (► Fig. 19.2). Traumatic wounds are not managed the same as sterile operative incisions because they are at greater risk for infection and unfavorable scarring.

19.3 Nonoperative Treatment

19.3.1 Lacerations

Partial-thickness wounds do not require repair and can be managed similarly to an abrasion. The area is washed twice daily with gentle soap and water followed by antibiotic ointment for a few days. Epithelialization usually takes 7 to 10 days. The area

can appear pink for several weeks and should be kept out of the sun for at least 12 months to avoid hyperpigmentation.

Heavily contaminated wounds can be allowed to heal secondarily; an open wound is a safe wound and is very unlikely to become infected (a treatment for an infected wound is to open it; ► Fig. 19.3). Human or animal bite puncture wounds inoculate deep tissues with bacteria; closure of these injuries (even with antibiotics) is at high risk for infection. Washing the areas regularly and letting them heal secondarily will significantly reduce the risk of infection. If the scars heal unfavorably, they can safely be revised at a later time.

19.3.2 Intravenous Extravasations

The scalp is a common site for intravenous catheter placement in the neonate, and extravasation can cause alopecia. Other intravenous access sites include the upper or lower extremities. Most extravasations involve small volumes of benign fluids that do not cause morbidity. In one series of 1,800 extravasations, only 2% of resulted in skin injury, and no child had a compartment syndrome. Catheters are located in the superficial venous system, above the level of the muscle fascia; the risk of injury to deep structures is low.

The most important maneuver to eliminate the extravasated fluid is elevation of the extremity. Application of ice or heat is contraindicated because they may exacerbate the injury. Both ice and heat have been shown to worsen tissue damage after the extravasation of chemotherapeutic agents. Heat can cause thermal injury and ice may result in ischemia from vasoconstriction. Sensation in the extravasated area may be impaired and thus the patient might not be able to appreciate pain from the application of heat or ice. In addition, young children or intubated patients are unable to verbalize discomfort from hot or cold packs. Thus, heat or ice may cause a “second hit,” converting a partial-thickness skin injury into a full-thickness wound.

Our institution does not advocate for the use of antidotes after extravasations. Hyaluronidase and phentolamine are the most commonly prescribed, but randomized prospective studies on their efficacy are not available. If an antidote is to be effective, it must be administered early. Unfortunately, most extravasations are not appreciated immediately and by the time plastic surgical consultation arrives, the antidote is ordered and available for administration, the substance has already diffused or its effects have worn off. For example, phentolamine has been used to treat epinephrine extravasation, but the effects of epinephrine wear off in 60 to 90 minutes, and the added volume of phentolamine may worsen tissue injury. Injection of an antidote increases the volume of fluid and worsens pressure necrosis.

Partial-thickness skin injuries are managed by topical antibiotic ointment; they typically heal without scarring by epithelialization in 7 to 10 days. Full-thickness injuries are managed with topical antibiotic ointment or dressings (► Fig. 19.4). Debridement is indicated for necrotic tissue to reduce the risk of infection and to facilitate healing by secondary intention. Following debridement, damp-to-dry saline dressing changes twice daily

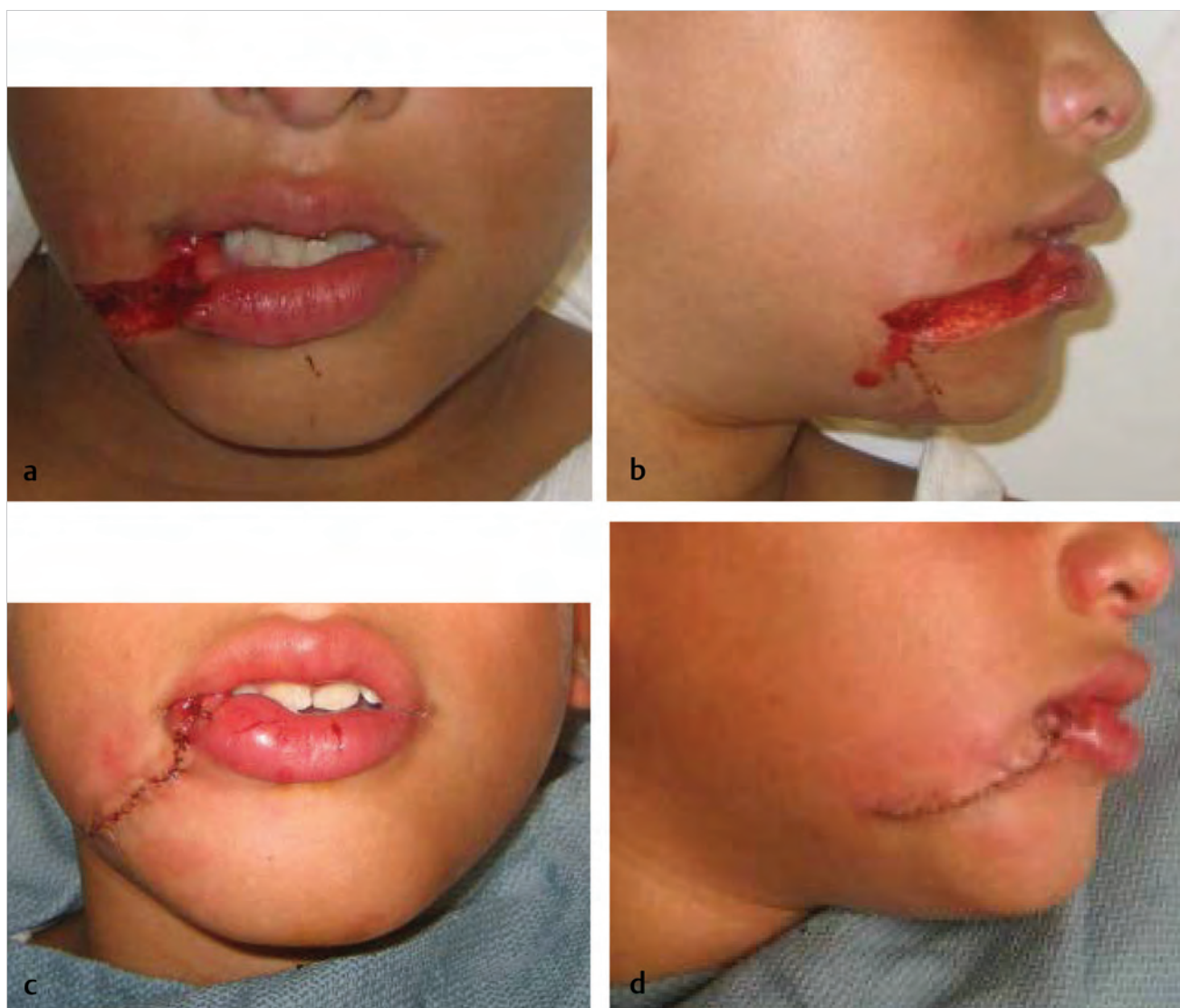


Fig. 19.1 Facial dog bite injury. (a,b) Preoperative appearance. (c,d) Following repair. Prior to closure, mucocutaneous junction landmarks were marked and approximated to best align the white roll.

are used. Full-thickness skin injuries are allowed to heal secondarily. If a compartment release is performed, the wound is managed with a vacuum-assisted wound closure device for 1 to 2 weeks as the swelling resolves and is then allowed to heal secondarily or a delayed primary closure is performed. Skin grafts should be avoided because there is no loss of skin and grafts cause a significant deformity.

19.4 Operative Treatment

19.4.1 Indications

Before deciding on closing a laceration, the risks of infection, sedation, and trauma to the patient/family must be considered. All traumatic wounds are contaminated and the risk of infection can be significant if they are closed. Heavily contaminated wounds (e.g., dog bite) should be closed cautiously. The goal of repairing large, problematic injuries is to minimize the wound

burden to the child while lowering the risk of infection as much as possible (► Fig. 19.5). If the wound becomes infected the child, who has already undergone a significant trauma and subsequent repair, must then undergo an incision and drainage followed by secondary healing and likely scar revision. In contrast, if small puncture wounds from a dog bite are allowed to heal secondarily, the child may only need a scar revision in the future.

19.4.2 Sedation

The decision to perform a laceration repair using a local anesthetic, sedation in the emergency room, or general anesthesia depends on the age of the patient and extent of the injury. The younger the child and larger the wound, the more likely it should be repaired under general anesthesia. Lacerations in young adolescents typically can be performed with local anesthesia. Most infants and children require sedation administered by the

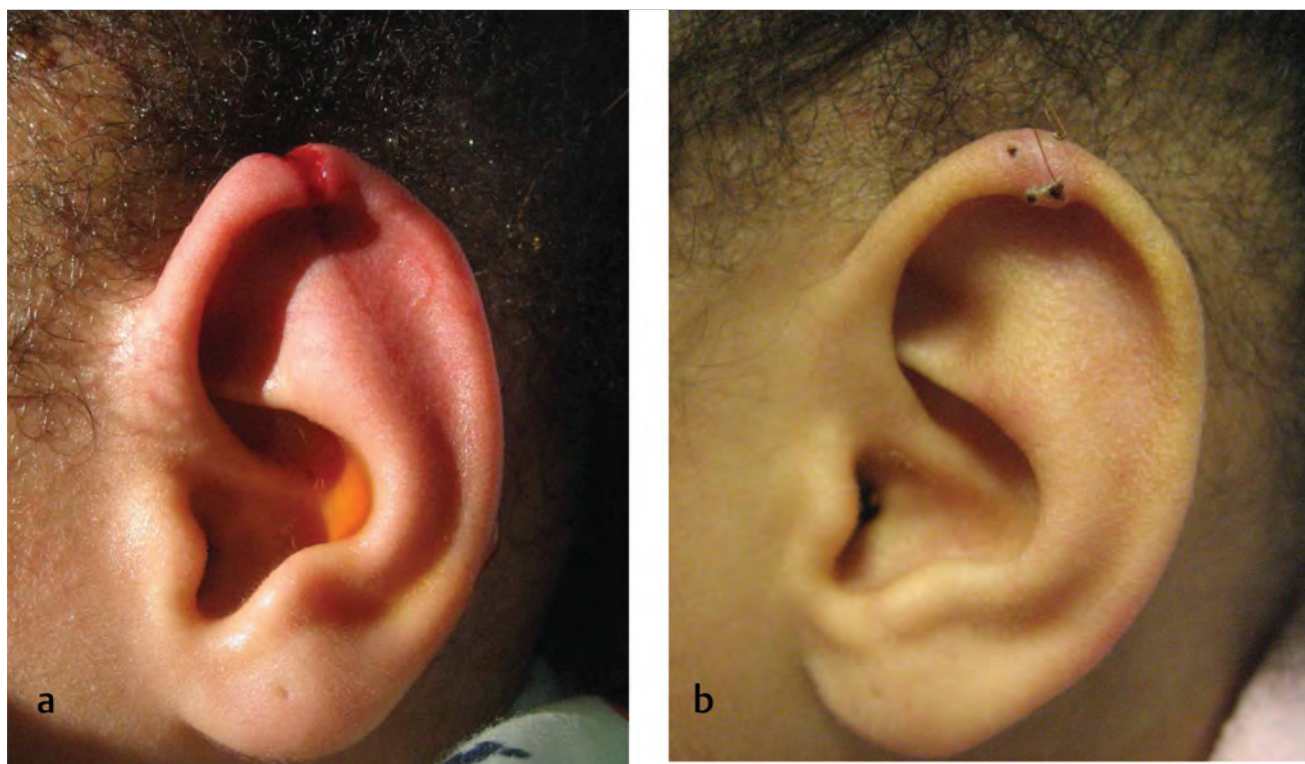


Fig. 19.2 Ear laceration in a child repaired with dissolvable chromic sutures. (a) Appearance of injury. (b) Three weeks postrepair, the child does not require suture removal.

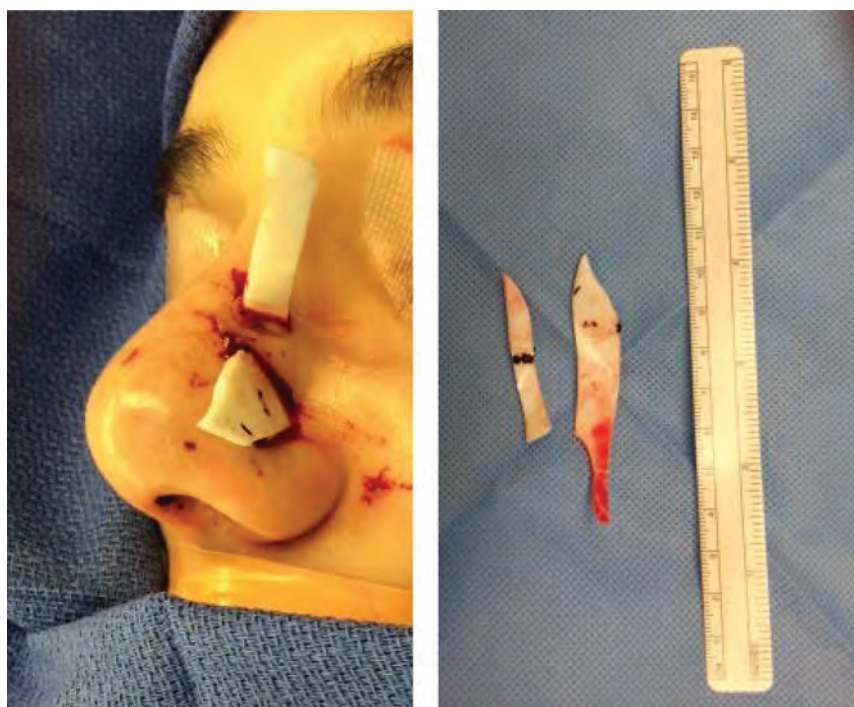


Fig. 19.3 Adolescent female who suffered an assault. Pieces of a dirty plastic bowl caused puncture wounds, entered the sinus, and lacerated mucosa. After removing the foreign bodies and irrigating the areas, the puncture sites were allowed to heal secondarily to reduce the risk of infection.

emergency room staff. Sedation in this age group gives the patient a better experience compared to local anesthesia, and facilitates the repair because the child is still. Any large injury (independent of the age of the patient) that involves significant time and effort to repair should be done under general anesthesia to ensure the patient has the most favorable outcome.

19.4.3 Antibiotics

Clear data regarding the use of antibiotics for traumatic soft-tissue injuries in children do not exist. Because traumatic wounds can be considered clean contaminated or contaminated, guidelines for operative incisions suggest that antibiotic prophylaxis



Fig. 19.4 A 10-month-old with full-thickness skin loss following intravenous extravasation. (a) Eschar. (b) Following separation of the eschar. (c) Seven weeks after the injury, the wound healed secondarily.



Fig. 19.5 An 8-year-old male who suffered a significant dog bite injury. (a) Preoperative appearance. (b) Following repair. The wound was closed loosely with interrupted, dissolvable sutures. A drain was placed to ensure that part of the wound remained open to reduce the risk of infection. (c) Healed sites 12 months following the injury. (d) Following revision of hypertrophic facial scars. The patient subsequently underwent another scar revision.



Fig. 19.6 Teenage male who suffered traumatic stab wounds to the face. Preoperative examination was consistent with division of the marginal mandibular branch of the facial nerve that was repaired at the same time as the skin lacerations. **(a)** Stab wound sites. **(b)** Divided facial nerve branch. **(c)** Following repair of the facial nerve injury.

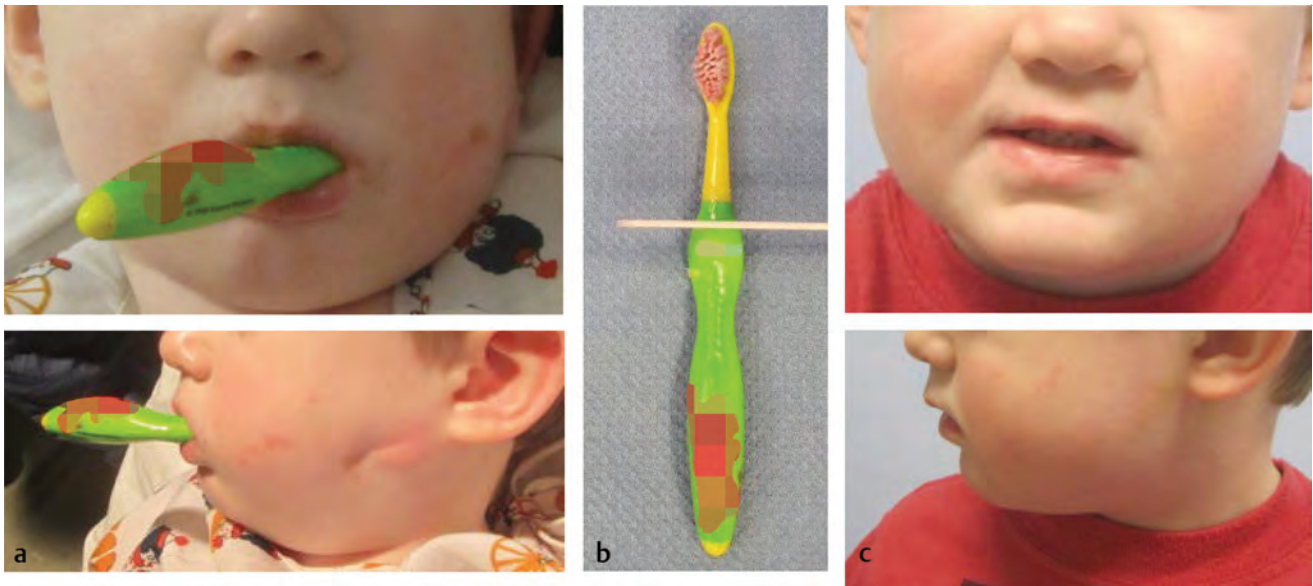


Fig. 19.7 A 3-year-old male fell off his bed with a toothbrush in his mouth that lacerated the oral mucosa and became embedded in his cheek. After removing the foreign body, the parotid duct was irrigated with saline to ensure its patency. **(a)** Preoperative appearance. **(b)** Removed toothbrush. The area distal to the wooden tongue blade was located in the cheek. **(c)** Postoperative appearance.

is indicated. My practice is to give one dose of antibiotics before wound closure in noncomplicated lacerations. I manage contaminated lacerations (e.g., dog bite, foreign body) with a preoperative dose of intravenous antibiotics followed by 5 to 7 days of oral antibiotics. Because a postoperative infection is a more significant problem in a child compared to an adult, I have a low threshold to administer antibiotics. An infection in children is more traumatic to the patient/family and incision and drainage may require sedation or general anesthesia.

19.4.4 Wound Inspection

The first step in laceration repair is to infuse local anesthetic containing epinephrine. The anesthetic eliminates pain while irrigating and the epinephrine facilitates the repair by reducing bleeding. Epinephrine can be used in any part of the body (e.g.,

nose, fingers, toes) and will not cause tissue ischemia because it only lasts 60 to 90 minutes. Without the use of epinephrine, the risk of iatrogenic injury and a less optimal repair is increased. After the infusion of local anesthetic, the wound is prepped and draped. Next, the area is inspected to determine the severity of the injury, necrotic tissues, and the presence of foreign substances. A foreign body should be removed prior to wound closure to reduce the risk of infection. However, if a material cannot be located and the risks of aggressive exploration outweigh the benefits of removing the substance, the wound can be closed over the material (plastic surgeons routinely place foreign materials throughout the body). Ideally, facial nerve transections are repaired at the time of the laceration repair (► Fig. 19.6). A laceration of the parotid duct is determined by cannulating it intraorally and injecting saline (► Fig. 19.7). Duct injuries are repaired over a stent (► Fig. 19.8).

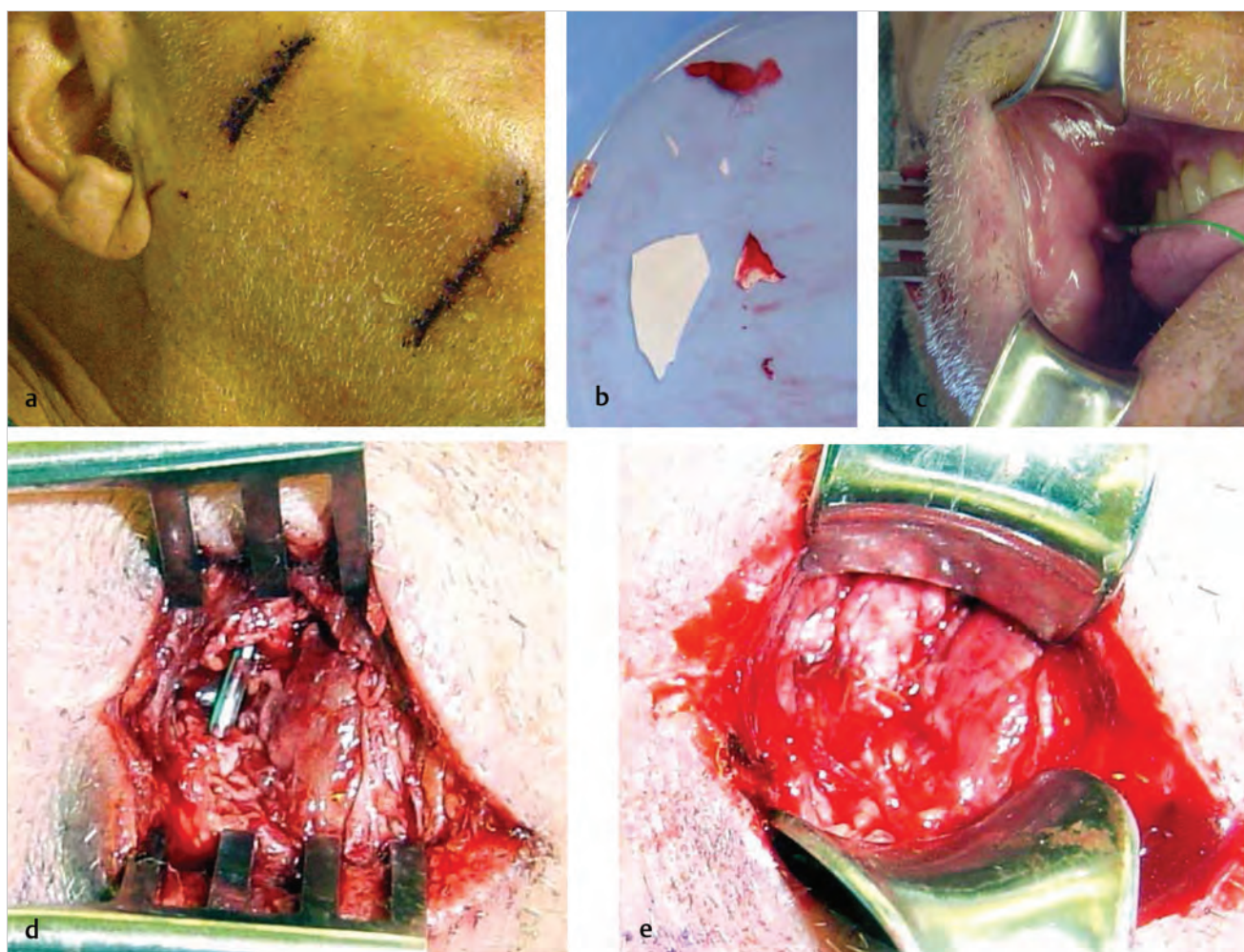


Fig. 19.8 Repair of a parotid duct injury. (a) Lacerations sustained when the patient fell into glass. (b) Glass removed from the cheek wounds. (c) Parotid duct cannulated and irrigated; saline in the cheek wound indicated a parotid duct laceration. (d) Stent located in parotid duct laceration. (e) Parotid duct repaired over a stent.

19.4.5 Irrigation and Debridement

An important step in laceration repair is ensuring the most favorable environment prior to wound closure. After the area has been inspected, it is irrigated to reduce the amount of bacteria and foreign material in the wound. The greater the contamination, the more the wound should be washed out. Irrigation is critical to reduce the risk of postrepair infection by converting a contaminated wound to a cleaner environment. Following irrigation, damaged skin edges that are ischemic or have traumatic tattooing should be debrided (► Fig. 19.7). Suturing fresh, noninjured skin edges will give the best possible scar and reduce the risk of infection and wound dehiscence.

19.4.6 Wound Repair

Complicated reconstructions at the time of injury should be avoided. Wounds are closed linearly or allowed to heal secondarily. Placing a skin graft or doing a local-regional free tissue transfer has a higher risk of failure and complications compared to reconstruction of a healed/stable area (► Fig. 19.8). Traumatic wounds are closed differently than sterile operative incisions. A

major principle is that the wound should not be closed tightly or occluded. Instead, the laceration is closed loosely and allowed to drain to reduce the risk of infection (► Fig. 19.9). Generally, I place one-half of the amount of sutures that I would use to close a sterile operative incision. For the intradermal layer I prefer Vicryl because it handles better than Monocryl and is less likely to unravel. To close the epidermis and superficial dermis, I typically use interrupted chromic in children and nylon in adolescents.

Placing nylon sutures in children should be avoided because (1) removing them will be stressful to the child and parents who have already suffered a traumatic experience (laceration and repair) and (2) the likelihood of iatrogenic wound dehiscence is high when removing sutures in an uncooperative child. For facial lesions in children, we use 7-0 chromic, which lasts 3 weeks until the wound strength is approximately 20%; suture marks do not occur because the diameter of 7-0 approximates that of pore size. A general rule is that if sedation was required to place sutures, then the sutures should be absorbable because removing them will likely require sedation as well.

Fast-absorbing plain gut of 6-0 should not be used because although the suture dissolves in 5 to 7 days, it causes significant



Fig. 19.9 Dog bite injury to the face with ischemic wound edges. (a) Initial appearance. (b) After irrigation and debridement of damaged skin edges. (c) Loose closure over a drain.



Fig. 19.10 Adolescent female who suffered a dog bite injury to the lips. (a) Significant loss of tissue. (b) After irrigation and linear closure of lacerations. Remaining open areas were allowed to heal secondarily. An attempt was not made to reconstruct the lips with grafts and/or flaps. (c) Appearance 12 months postoperatively. She subsequently underwent scar revision and lip augmentation using dermal-fat grafting.

inflammation and leaves visible suture marks on the face. Also, after the suture dissolves, the wound only has 5% strength and the scar is likely to widen. Continuous sutures should not be placed in traumatic wounds because they prevent drainage and increase the risk of infection. Similarly, cyanoacrylate glue is avoided because it seals the wound, prevents drainage, and increases the risk of infection (► Fig. 19.10). Cyanoacrylate glue only provides strength for 7 to 10 days and the scar is likely to spread after the glue has dissolved.

19.4.7 Dressing

Nonocclusive dressings should be applied to traumatic wounds to allow drainage. On the face topical antibiotic, steri-strips, or gauze with tape can be applied. For significantly contaminated wounds, a Penrose drain can be placed to ensure adequate wound drainage. The parents remove the drain in 48 hours at home. Drainage from the site usually continues for another 2 to 3 days until the wound edges become approximated. Generally, the laceration can be washed with soap and water in the shower beginning 24 hours after the injury. Cleaning the area removes dried blood favorable for bacteria, and gives the most favorable environment for scarring.

19.4.8 Postoperative Management

Patients who have suffered human or animal bite wounds are examined 3 to 5 days after the injury to ensure that an early

infection is not occurring. Noncomplicated injuries are re-examined 3 weeks following the injury. At the postoperative appointment, steri-strips are placed over the scar and the parents are instructed to replace them for 3 weeks when the wound has achieved its maximal strength. Placing steri-strips prevents the scar from widening, protects the area from sun exposure, and reduces the risk of the accidental injury from the child. The parents are instructed to place sunblock and protective clothing for 12 months to prevent hyperpigmentation of the scar. I do not favor scar products for normal scars that are not hypertrophic/keloid. Patients are instructed to return 1 year following the injury if they are unhappy with the appearance of the scar. At this time, a decision can be made to determine if a scar revision should be done under sterile, nontraumatic conditions.

19.5 Complications

The most common acute problems following soft-tissue injury repair are infection and wound dehiscence. Early infections with cellulitis may be treated with oral antibiotics. If purulent drainage is present, then the wound should be partially or completely opened, depending on the severity of the infection and length of the wound. Generally, gauze is placed for 2 days to ensure drainage and prevent reapproximation of the wound edges. After 2 days, the gauze is removed and the wound is washed twice a day and allowed to heal secondarily. Severe infections may require intravenous antibiotic therapy.



Fig. 19.11 A 6-year-old male who suffered a dog bite injury to the cheek. (a) Initial appearance. (b) The area was closed loosely to reduce the risk of infection. (c) Cheek scar 12 months postoperatively. (d) Improved appearance following scar revision.



Fig. 19.12 Dog bite injury to the cheek closed at an outside hospital using cyanoacrylate glue. He presented 3 days later with a wound infection that was managed by incision, drainage, and intravenous antibiotics.

If a laceration closure dehisces, then it should be washed twice a day and allowed to heal secondarily. Delayed primary closure should not be performed because it is better for the scar to fully mature and perform a scar revision if necessary. Revising the scar will give a more favorable long-term outcome compared to delayed primary closure in a nonsterile, inflammatory environment.

The most common long-term problem following the repair of a soft-tissue injury is an unfavorable-appearing scar. Because a

traumatic wound has a greater bacterial burden, inflammation, and injured wound edges, the scar is more likely to be worse compared to closure of an operative incision. Decisions regarding whether or not a scar may be improved with scar revision should be made after it has fully matured 12 to 18 months following the injury. Often a scar will improve significantly over the first year and patients will avoid having an unnecessary procedure.

19.6 Conclusion

Pediatric plastic surgeons commonly manage soft-tissue injuries, particularly facial lacerations. Soft-tissue trauma in children is managed differently than in adults or sterile operative incisions. There should be a low threshold to repair injuries using sedation/general anesthesia and to administer antibiotics. Absorbable sutures are used whenever possible to avoid suture removal, which is another traumatic experience for the child.

19.7 Key Points

- Because lacerations are not clean wounds, they are managed differently than a sterile operative incision.
- An open wound is a safe wound; allowing heavily contaminated wounds to heal secondarily should be considered.
- Most pediatric patients require sedation or general anesthesia to repair their wound.
- Absorbable sutures should be used when possible.
- Intravenous extravasation injuries have a very low risk of compartment syndrome (unless it is a chemotherapeutic agent) and definitive management is elevation.

Suggested Readings

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20 Ear Reconstruction

Arin K. Greene

Summary

Ear deformities are common in the pediatric population. Disorders include: constricted ears, microtia, accessory cartilage, lobule anomalies, prominent ears, cryptotia, Stahl deformity, keloids, and traumatic defects. The reconstructive procedure and timing depends on the type of deformity.

Keywords: ear, reconstruction, otoplasty, cryptotia, Stahl, keloid, pediatric

20.1 Introduction

Several types of ear anomalies affect the pediatric population. These are reconstructed using different otoplasty strategies. Construction of an absent ear (microtia) is presented in a separate chapter. The primary morbidity of an ear anomaly is psychosocial. Children can develop lowered self-esteem if peers notice their malformation. Ear anomalies are not painful and reconstruction does affect hearing; consequently, insurance companies may not cover the procedure. Correction of some deformities (e.g., cryptotia) can facilitate the use of eyeglasses. Ear anomalies may be broadly divided into three categories: (1) excess structures, (2) deficient structures, and (3) structures present but malformed.

In contrast to most other types of congenital anomalies managed by pediatric plastic surgeons, ear malformations can cause increased anxiety for families because (1) they often have to

pay for the procedure and (2) they are subjecting their child to general anesthesia for a “cosmetic” problem. Consequently, surgeons treating ear anomalies should be well trained on the subject and ensure that patients and families have reasonable expectations (similar to rhinoplasty). For example, I emphasize that the goal of the operation is to improve the child’s appearance, but that I “guarantee” there will be some residual asymmetry between the ears.

20.2 Diagnosis

Diagnosis of an ear anomaly is made by physical examination (► Fig. 20.1, ► Fig. 20.2). The surgeon should be able to identify the abnormality and list reconstructive options. A framework for the operative approach usually can be made by determining if structures need to be removed, added, or rearranged. Ear anomalies can be better understood by knowing the embryology of ear development. The first mandibular arch gives three anterior hillocks: (1) tragus, (2) root of helix, and (3) superior helix. The second hyoid arch results in three posterior hillocks: (1) antihelix, (2) antitragus, and (3) lobule. The external auditory meatus is formed by the first branchial groove (► Fig. 20.3).

The surgeon must understand normal ear anatomy and can use a contralateral normal ear, if present, as a guide (► Fig. 20.4). The axis of the ear is angled approximately 20 degrees more posterior than a vertical line. The top of the helix is at the level of the eyebrow and the lobule is at the base of the columella. The height of the ear is 6.5 (± 1) cm and its width is

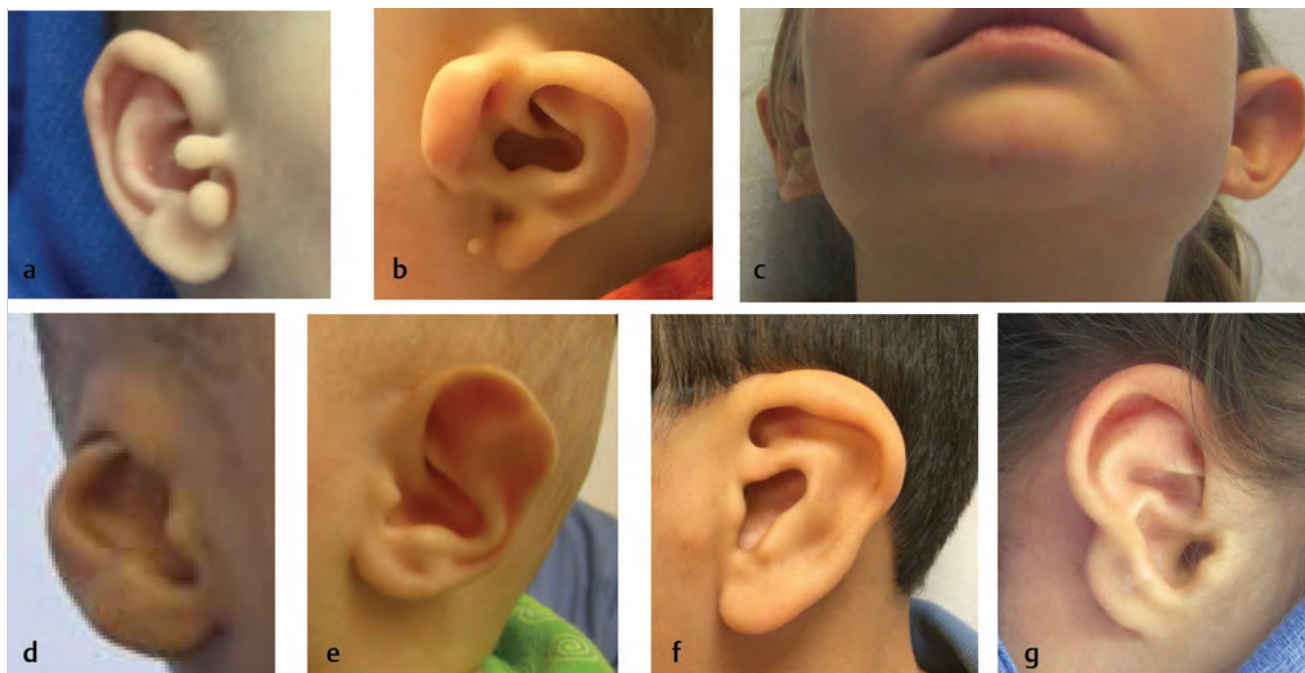


Fig. 20.1 Types of ear anomalies. (a,b) Accessory cartilage. (c) Prominent ear. (d) Cryptotia. (e) Stahl deformity. (f) Abnormal helical root and antihelix. (g) Lobule abnormality.

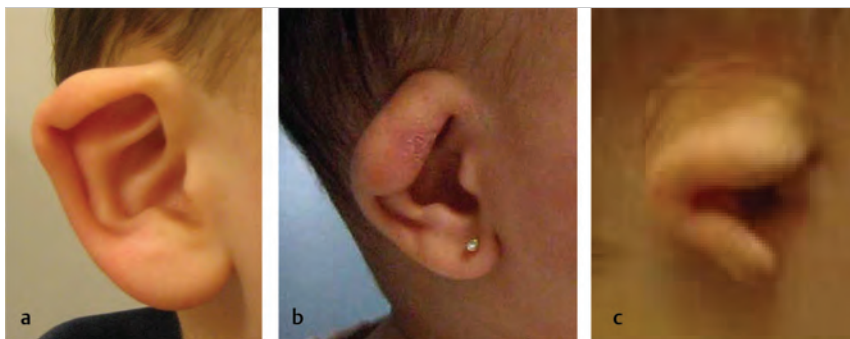


Fig. 20.2 Examples of a constricted ear. (a) Mild. (b) Moderate. (c) Severe.

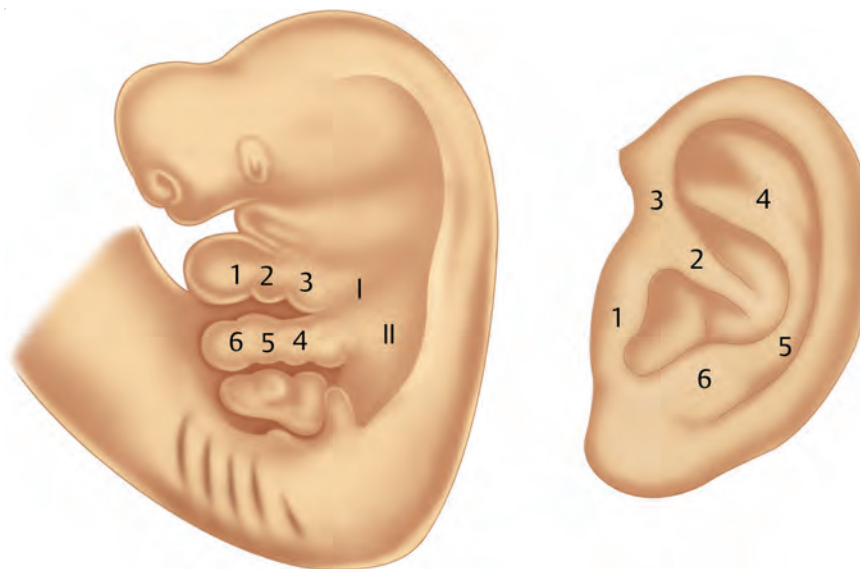


Fig. 20.3 Ear development. Six hillocks from the first and second arch form the ear.

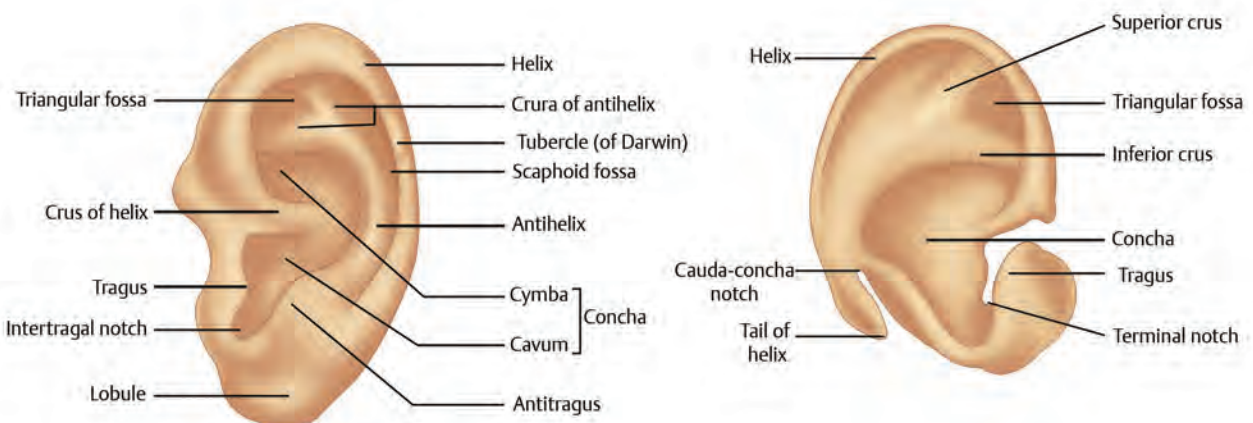


Fig. 20.4 Anatomy of the ear.



Fig. 20.5 Types of ear molding. Above: commercially available system. (a) Device placed around ear. (b) Piece molds the helix. (c) Conchal bowl conformer to flatten the root of the helix. (d) Cover applied. Below: custom wire, mold, and tape. (e) Anomaly of the helix. (f) Wire. (g) Mold and tape. (h) Improved appearance of the ear.

4.0 (± 0.5 cm). It is located 6 to 7 cm posterior to the lateral canthus. Although several measurements for normal protrusion from the mastoid have been described, I use 15 mm (top helix), 20 mm (midhelix), and 22 mm (lobule).

Imaging is rarely necessary to manage an ear deformity. If a branchial cleft cyst or sinus is suspected, however, I will obtain a magnetic resonance imaging to confirm the diagnosis and determine the extent of the tract. Occasionally, a sinus can be deep and approximate the facial nerve; this information is helpful prior to operative intervention.

20.3 Nonoperative Treatment

A window exists up to 3 months of age when the ear can be molded. Because one-third of anomalies improve during the first 2 weeks of age (particularly a prominent helical root), molding should not be initiated until after this time. Ideally, molding is started between 2 and 4 weeks of age when maternal estrogens in the child facilitate cartilage manipulation. The ear can be molded using wire and tape or a commercially available system (► Fig. 20.5). Both methods are equally effective; the commercial system is easier to use, but more expensive.

Unfortunately, patients often are referred after the window to mold their ear has passed. I will not attempt to mold the ear after 3 months of age because of the poor response. The ear deformity that is most amenable to molding is a prominent ear (► Fig. 20.6). The antihelical fold is straightforward to create and the concha is able to be set back. Another anomaly that is able to be molded includes a mild constricted ear with

overhanging helical cartilage; the cartilage can be “lifted” into better alignment. A Stahl deformity also can be improved by flattening the abnormal crus. Children commonly are referred for ear anomalies that cannot be helped with molding (e.g., severe constricted ear, cryptotia).

20.4 Operative Treatment

20.4.1 Indications

The primary indication for correction of an ear anomaly is to improve the patient’s self-esteem. Cryptotia and moderate-severe constricted ears can make it difficult to wear eyeglasses. Timing of intervention falls into three categories: (1) 6 to 12 months, (2) 3 to 4 years, (3) late childhood or adolescence. Simple excisions of excess structures can be performed during infancy under local anesthesia. Parents with preauricular cartilage remnants are usually eager to remove them as soon as possible. Most are small and able to be excised under local anesthesia in the office. I prefer that he or she is at least 6 months of age, when their physiology approximates that of an adult, before doing an elective procedure. After 12 months of age, however, it is too difficult to restrain an awake child and I will perform the procedure in the operating room with sedation or wait until the child is old enough to be able to cooperate in the clinic. If a child has an ear deformity that cannot be corrected simply using local anesthesia during infancy, I prefer they be at least 3 years of age before operative intervention under general anesthesia.



Fig. 20.6 Examples of ear molding. (a,b) Prominent ear. (c,d) Constricted ear. (e,f) Stahl ear.

Because long-term memory and self-esteem begin to form at approximately 4 years of age, many parents decide to correct deficient or malformed structures when the child is 3 years of age. At this time, the ear has achieved 85% of its size and the risk of scar limiting enlargement of the ear is reduced (ear growth is complete by approximately 6 years of age). Although studies have shown that operating on ears before 3 years of age does not inhibit growth, I prefer to wait until at least 3 years of age because (1) the ear is larger, which facilitates the procedure, and (2) it is not urgent to intervene before this time because memory and self-esteem have not yet formed.

If a child has a severe deformity that he or she will definitely want improved, then it is best to do the procedure prior to 4 years of age, before memory and self-esteem form. Children at this age tolerate the procedure more favorably than older patients who are more anxious about the operation. If an individual has a mild deformity that he or she may not want to

have improved, then it is prudent to wait until they are older to determine if they become bothered by it.

Some families choose to wait to correct an ear anomaly until the child is old enough to participate in the decision to have the procedure. Children between 5 and 8 years of age typically are bothered by a deformity, but fear of the operation outweighs their desire to improve their appearance. After 8 years of age, the child's interest in correcting the anomaly begins to outweigh the anxiety about the operation. Girls typically present later than boys because they are better able to camouflage their deformity with hair. Boys often will grow their hair long to hide their ear.

Except for small excisional procedures in infancy, I prefer to correct other ear anomalies under general anesthesia. Even in adolescents who may tolerate procedures under local anesthesia, they typically have a better experience if they are asleep. General anesthesia also facilitates the procedure for the surgeon and increases the likelihood of achieving the most favorable outcome.

During any ear procedure, I ensure that betadine has been placed into the external auditory meatus. I do not administer a perioperative dose of antibiotic unless I am (1) doing an extensive cartilage dissection or (2) placing permanent intracartilaginous sutures. Although these procedures are clean cases, the benefit of reducing the risk of infection, chondritis, and destruction of the ear after a "cosmetic" operation outweighs the potential disadvantages of the antibiotic. I do not prescribe postoperative antibiotics.

The ears are particularly tolerant of asymmetry because of their lateral position over the temporal bones. The full appearance of the ears cannot be appreciated on frontal view. To compare the ears, an individual must look at one ear first and then change positions to see the contralateral side. Consequently, it is most important to have the ears as symmetrical as possible on frontal view. Because most ear anomalies cause a "cosmetic problem," patients and families can be focused on minor asymmetries. It is important that families understand that slight asymmetries between the ears are common in the general population and that there will be residual asymmetry following the procedure.

20.5 Types of Abnormalities (Listed in Order of Frequency)

20.5.1 Accessory Cartilage

Accessory cartilage in the preauricular area is the most common ear anomaly (► Fig. 20.7). I do not advocate placing a suture around the area to cause necrosis because (1) there will be a residual bump that has a high likelihood of needing a surgical procedure to improve and (2) restraining the child to perform a definitive excision is not much more involved than tying a suture around the lesion. I prefer to remove the accessory cartilage between 6 and 12 months of age under local anesthesia in the office to obviate general anesthesia. Patients who present after 12 months of age are managed with sedation in the operating room because it is too difficult to restrain the child in the clinic. If an infant has a large lesion with a wide cartilage base that cannot be easily excised and closed, the procedure is performed in the operating room. Only dissolving 6–0 or 7–0

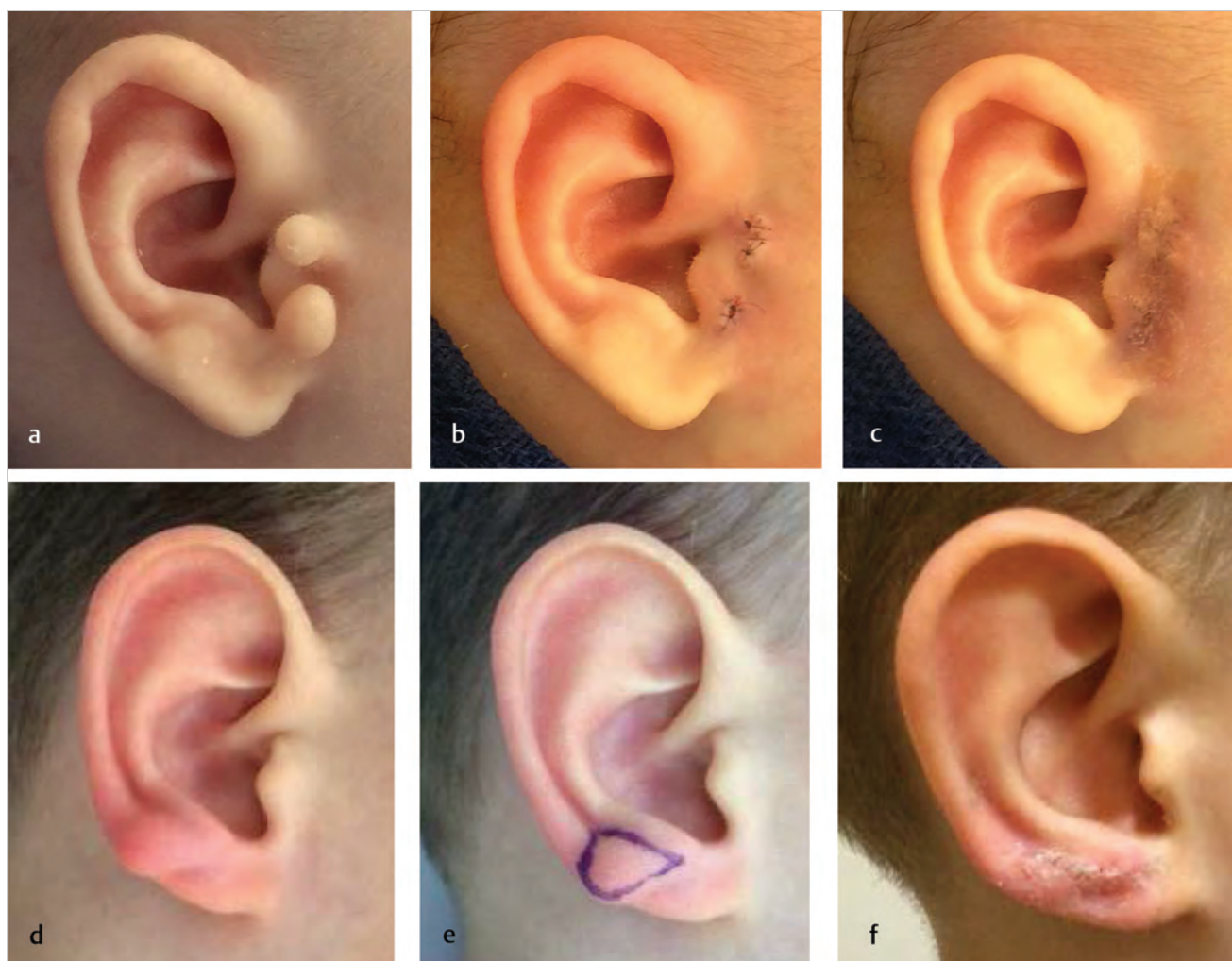


Fig. 20.7 Accessory cartilage. (a) Two preauricular lesions. (b) Following excision and repair with 7–0 chromic suture. (c) Cyanoacrylate glue and a steri-strip are placed to reinforce the area. (d) Accessory tissue on lobule. (e) Outline of resection. (f) Following removal.

chromic sutures are placed followed by cyanoacrylate glue and steri-strips.

20.5.2 Lobule Defects

Split Earlobe

This deformity results from an earring being pulled through the lobule (► Fig. 20.8). The cleft is rapidly epithelialized preventing the edges from re-approximating. Split earlobes in adolescents can be repaired with local anesthesia; children require general anesthesia. The cleft is de-epithelialized and approximated using absorbable sutures in a straight line. The end of the cleft is closed with a mattress suture for eversion to prevent a notch from scar contraction. A 5–0 Vicryl intradermal suture is used, followed by interrupted 7–0 chromic for the anterior surface of the ear and 6–0 chromic for the posterior area. The closure is protected with cyanoacrylate glue and steri-strips. The scar can be re-pierced 3 months later when it has achieved its maximal strength of 80% of noninjured tissue. The advantages of re-piercing along the scar is that the scar is camouflaged, and if the earring pulls through again, a second scar is not formed.

The disadvantage of piercing the scar is that it is not as strong as the adjacent lobule and thus has a higher risk of pulling through again. If families choose to re-pierce the scar, they should not use heavy earrings. My preference is to pierce the ear adjacent to the scar.

Keloid

The earlobe has a greater risk of forming keloid scars from piercings, compared to other parts of the body. Keloids are managed with pressure, corticosteroid injection, and/or resection. Small, superficial lesions can be treated with pressure only. Patients are advised to wear a clip-on earring or purchase a pressure earring designed for keloids. The pressure device is worn only at night when sleeping. A pressure earring is a good first-line intervention for keloids in young children who are less tolerant of undergoing a corticosteroid injection in the clinic. Larger lesions can be managed with serial triamcinolone injections in addition to the pressure earring. I prefer 10 mg/ml rather than 40 mg/ml to reduce the risk of fat atrophy. A 30-gauge needle and 1-mL syringe is used. The corticosteroid lasts approximately 2 weeks and patients return every 3 to 4 weeks.

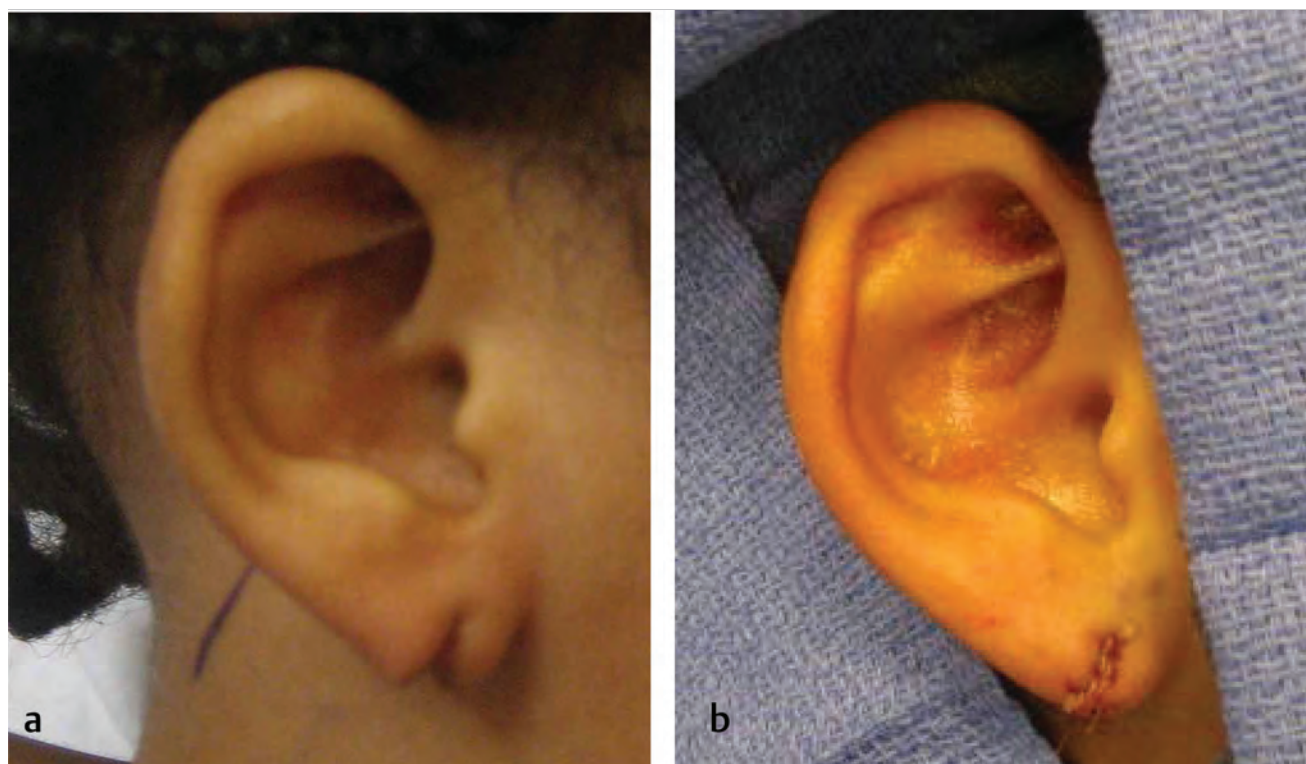


Fig. 20.8 Split earlobe deformity. (a) Preoperative appearance. (b) Following repair.

for repeat injections until the desired result is obtained. Large keloids are best managed with resection because pressure/injections will only give minimal improvement and the patient will continue to have a significant deformity. Families are educated that resection is exponentially more traumatic to the ear than the piercing that caused the keloid. Consequently, without close follow-up and the use of postoperative compression and injections, the keloid will return and can be worse. After resecting the keloid, I inject triamcinolone at the end of the procedure. Dissolvable sutures are used and the patient wears a pressure earring immediately at night. Four weeks postoperatively, the patient returns for monthly corticosteroid injections. After 6 months, the injections are discontinued and the patient continues to wear the nightly pressure earring for another 6 months until the scar has matured. Using this regimen, I have not had a keloid recur and I counsel patients that the original size of the keloid can be reduced by at least 80%. Although radiation has been described as a treatment for recurrent keloids, I have not had a patient require this intervention.

Expanded Lobule

Recently, ear gauges that expand the lobule have become popular. Patients who discontinue the gauge may present for lobule reconstruction. The earlobe is expanded in both a vertical and a horizontal plane. In order to reduce the size of the lobule in two planes, I have reconstructed the defect by first de-epithelializing the area and then closing it using a purse-string suture or by removing horizontal triangles to lift the lobule, leaving the patient with a vertical and horizontal scar (► Fig. 20.9).

Congenital Deficiency

The most common lobule deficiency is a cleft that causes a deformity and obviates the ability to place earrings (► Fig. 20.10). The cleft is de-epithelialized and corrected by advancing the tissue on both sides to form a lobule. Rarely, a patient can have an absent lobule, on the spectrum of microtia. The lobule can be constructed using staged skin flaps from the adjacent neck, or by placing a conchal cartilage graft. I have reconstructed the lobule using both methods and found that using cartilage results in the most favorable outcome (► Fig. 20.11).

20.5.3 Sinus Tracts

Auricular sinus tracts are a common ear anomaly. They may express keratin and can become infected. Sinus tracts cause minimal deformity and do not mandate excision. Patients are instructed to massage the area once a day with a warm compress to express keratin and prevent the tract from being obstructed. Excision of sinus tracts should only be considered if a patient has repeated infections and has failed warm compresses. Resection of a sinus tract is at risk for recurrence, infection, and nerve injury if the tract extends significantly.

20.5.4 Prominent Ear

Approximately 5% of the population has a prominent ear caused by an (1) absent or weak antihelical fold and/or (2) overgrown conchal bowl. Most patients will require strengthening of the antihelix and setback of the concha. Rarely, a patient will have a normal antihelix and only require a procedure on the concha.

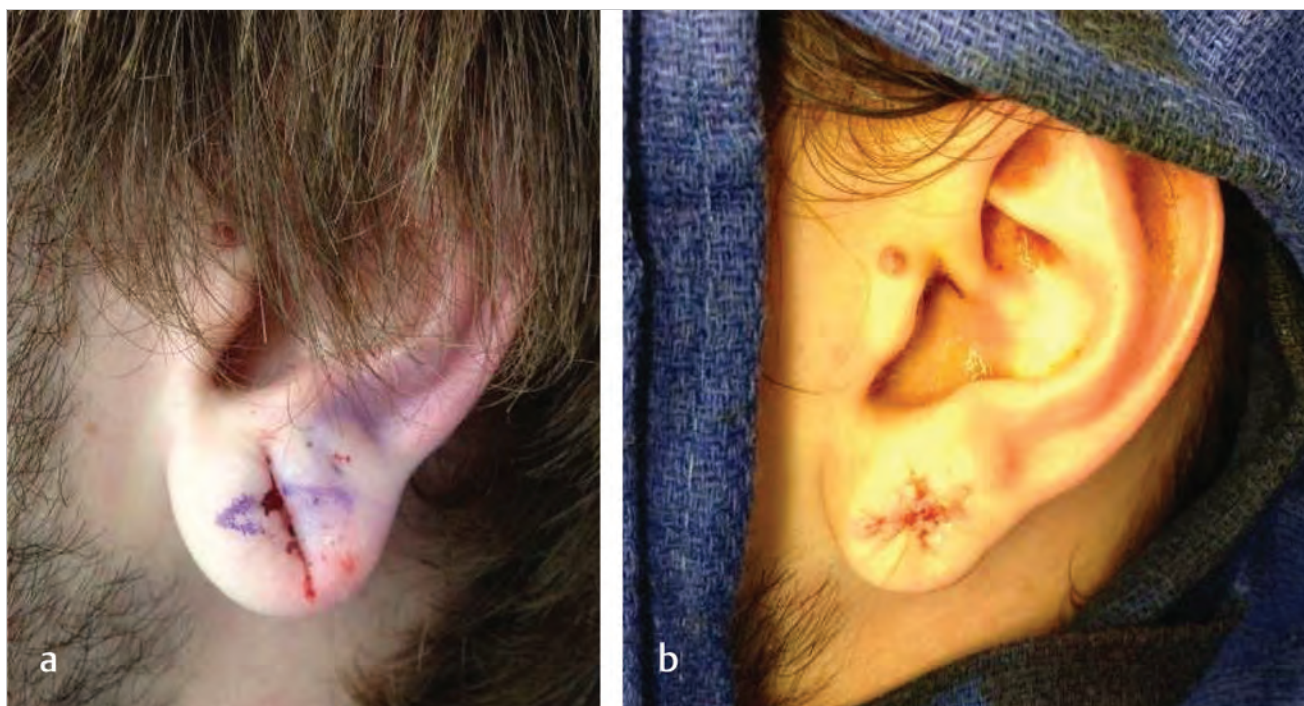


Fig. 20.9 Lobule deformity from an ear gauge. (a) Preoperative appearance with outline of horizontal wedge resection. (b) Following repair.

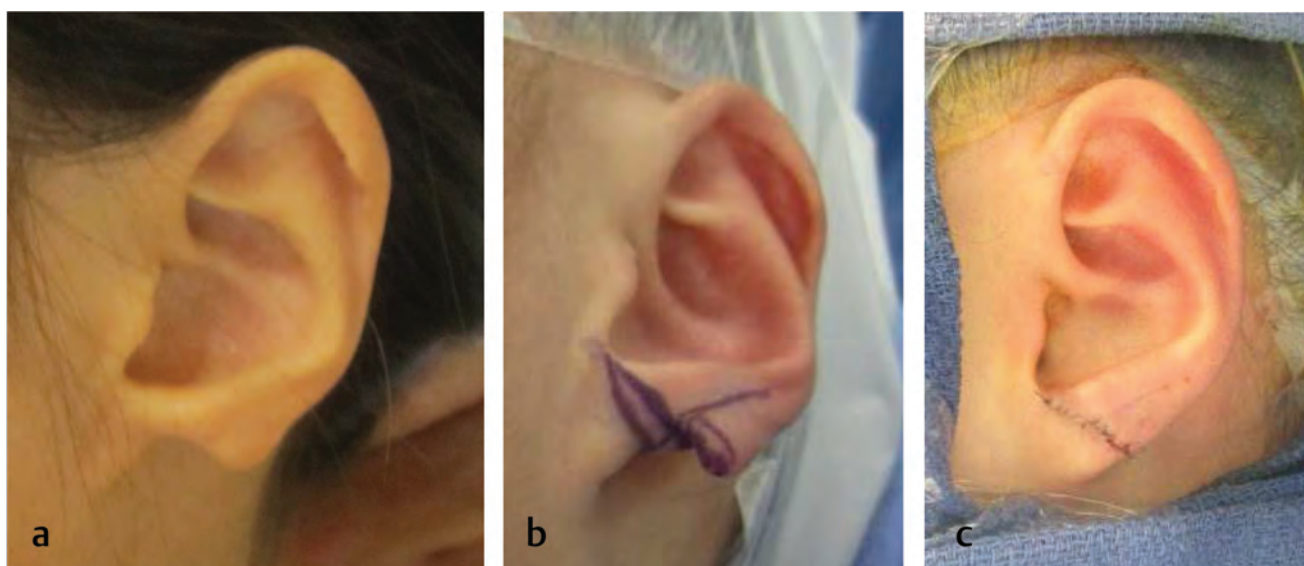


Fig. 20.10 Congenital lobule deficiency. (a) Preoperative view. (b) Outline of bilateral flaps. (c) Following advancement and rearrangement of tissue.

Similarly, a child may have a normal concha and only require creation of the antihelix. Although many definitions of a prominent ear exist, generally an ear will appear prominent if its superior or middle aspect is greater than 20 mm from the mastoid.

Although many techniques have been described to correct a prominent ear, I prefer (1) suture-only strengthening of the antihelix (Mustarde's technique: scapha-concha sutures) and (2) concha excision. Cartilage scoring (Stenstrom's technique) for the antihelical fold increases the complexity of the operation, is less predictable than suture placement, and is associated

with contour abnormalities. In the pediatric population, the cartilage is easily positioned using sutures and does not require weakening. To set back the middle third of the ear, I perform cartilage resection instead of concha-mastoid (Furnas) sutures. Suturing the hard cartilage to the weak mastoid fascia/periosteum has a high likelihood of suture dehiscence, stretching, and recurrence of the prominent ear. In contrast, removing cartilage from the concha ensures that the middle third of the ear is reduced and eliminates the possibility of recurrence.

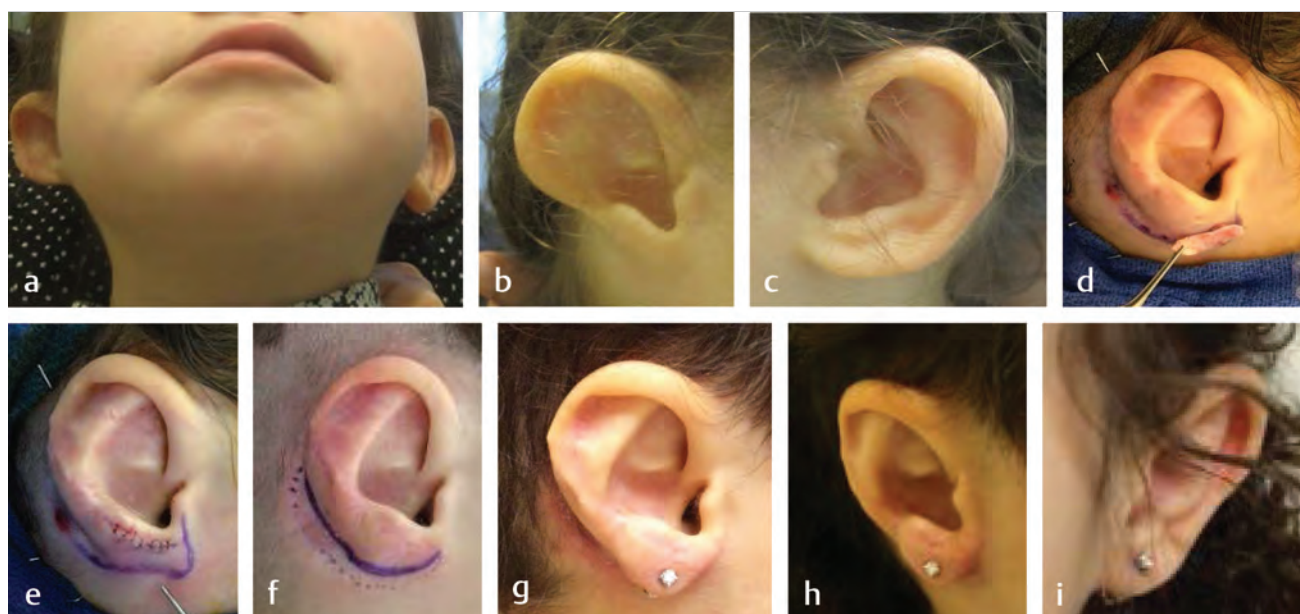


Fig. 20.11 Absence of lobule and prominent ear. (a,b) Preoperative appearance. (c) Normal contralateral ear. (d) Correction of ear prominence by strengthening the antihelix. Conchal cartilage is used to re-create the lobule. (e) After subcutaneous placement of the cartilage. Tip of instrument shows inferior position of the graft. (f) Outline of staged elevation of the soft-tissue and cartilage graft. The solid line marks the inferior aspect of the cartilage and the dashed line is skin that will be turned over to cover part of the posterior surface. An advancement flap and skin graft were used to re-create the retro-lobule sulcus. (g,h) Postoperative appearance. (i) Normal contralateral ear.

After the patient is under general anesthesia, I measure the distance of the ears to the mastoid from the most prominent part of the superior helix and from the midhelix at the level of the root. The ear is folded and the position of the antihelical fold is marked. Local anesthetic is infiltrated to hydrodissect the anterior skin off the cartilage to facilitate suture placement. A posterior skin excision is performed to facilitate the procedure by enhancing exposure. Four 4-0 permanent mattress sutures (mersilene) are placed along the antihelical fold. It is important not to overcorrect the antihelical fold, which should have a curved, soft appearance. On frontal view, the helix should be visible beyond the antihelix. An approximately 2- to 3-cm piece of superior conchal cartilage is removed. The width of the area should be 5 to 7 mm. The cartilage defect is repaired with interrupted 4-0 PDS suture. After the cartilage excision, a line of excess skin may be present anteriorly, but this improves over the next 12 months. If more than a 7-mm width of cartilage is removed, then it is possible that the excess skin anteriorly might be bothersome to the patient and require excision. Some surgeons perform an anterior skin excision at the same time as the posterior concha resection, but this places a scar on the front of the ear and if cartilage greater than or equal to 7-mm width is removed, the patient is at minimal risk for needing excess skin removed.

To set back the lobule, one Mersilene suture is placed through the tail of the helix and is sutured to the base of the conchal

bowl. A posterior lobule skin excision also is performed. The concha resection further helps to reduce the prominence of the lobule. Before closing the incision, I ensure that the helix appears straight, is not curved, and that a telephone deformity is not present. I also perform repeat measurements; the superior distance of the helix to the mastoid must be 15 (± 3) mm and the middle distance 18 (± 3) mm. These measurements should have a difference of ≤ 3 mm between the ears to ensure adequate symmetry. The incision is closed using interrupted 5-0 Vicryl suture followed by a running 6-0 chromic suture. Cyanoacrylate glue and a steri-strip are placed.

Postoperatively, I use a headwrap (large piece of cotton over the ear followed by a gauze wrap) that is worn for ≤ 1 week until the first clinic appointment. I do not attempt to place pressure on the ear with bolster sutures or pieces of cotton soaked in mineral oil. Patients then are instructed to wear a soft headband at night for 3 months postoperatively to prevent accidental anterior displacement of the ear that could dehiscence the antihelical sutures causing recurrent prominence. Twelve weeks postoperatively, scar tissue is strong enough to maintain the shape of the ear (\blacktriangleright Fig. 20.12).

20.5.5 Constricted Ear

A wide spectrum of constricted (e.g., “lop,” “cup”) ear anomalies exist, generally consisting of deficient structures. They can



Fig. 20.12 Correction of a prominent ear deformity. Above: unilateral anomaly. (a) Preoperative view. (b) Postoperative appearance. (c,d) Bilateral prominent ears.

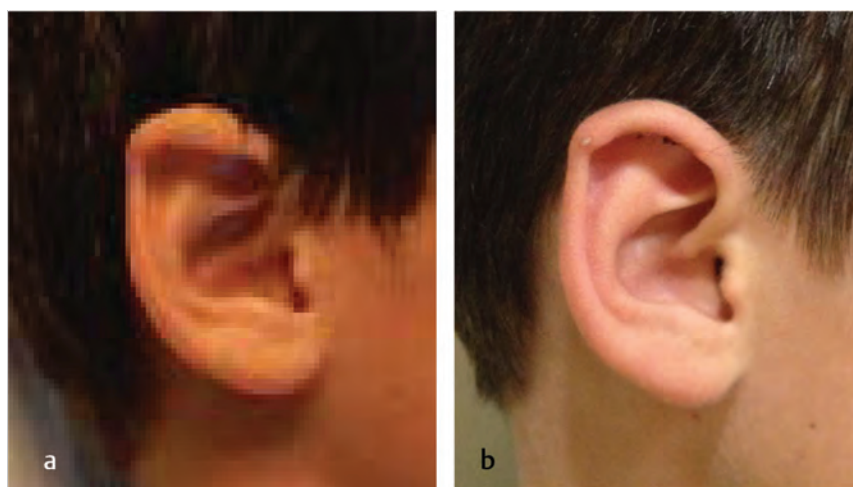


Fig. 20.13 Correction of mild constricted ear. (a) Preoperative image shows abnormal helix covering a portion of the superior crus of the antihelix. (b) Improved appearance following excision of cartilage.

range from minor helical problems to a variant of microtia and consist of (1) lidding of the helix, (2) prominence, and/or (3) reduced size from deficient structures. If possible, I prefer to manage these deformities as low on the reconstructive ladder as possible before considering cartilage grafts and retroauricular flaps. Minor deformities of the helix can be improved with simple excisions (► Fig. 20.13, ► Fig. 20.14). Moderately constricted ears that contain a sufficient amount of structures may be improved using techniques employed for prominent ear deformities (► Fig. 20.15, ► Fig. 20.16). Generally, the primary morbidity of a constricted ear is that it is prominent. If the ear is set back by strengthening the antihelical fold and placing sutures from the cartilage to the mastoid fascia/periosteum to lengthen the vertical dimension of ear, the appearance of the child can be significantly improved. Excision of overhanging helix also may be performed. Severely constricted ears that

approximate conchal-type microtia require total auricular construction using costal cartilage.

20.5.6 Cryptotia

Patients with cryptotia have a superior helix that is attached to the postauricular skin; a sulcus is not present. In addition to causing a deformity, patients have difficulty wearing eyeglasses. Many techniques have been described to reconstruct the anomaly, including local flaps and skin grafts. In my opinion, the most straightforward method to release the ear and provide a sulcus is to perform a local flap. I prefer using a one-stage V-Y advancement flap, which leaves a scar behind the ear and in the hairline (► Fig. 20.17). To protect the flap, I place a head dressing for 1 week.



Fig. 20.14 Treatment of a moderate constricted ear. (a) Preoperative image shows a wide helix obstructing the antihelix, scaphoid fossa, and triangular fossa. (b) Outline of the skin and cartilage resection. (c) Intraoperative view of the abnormal cartilage. (d) Following resection. (e) Skin closure along helix. (f) Two months postoperatively.

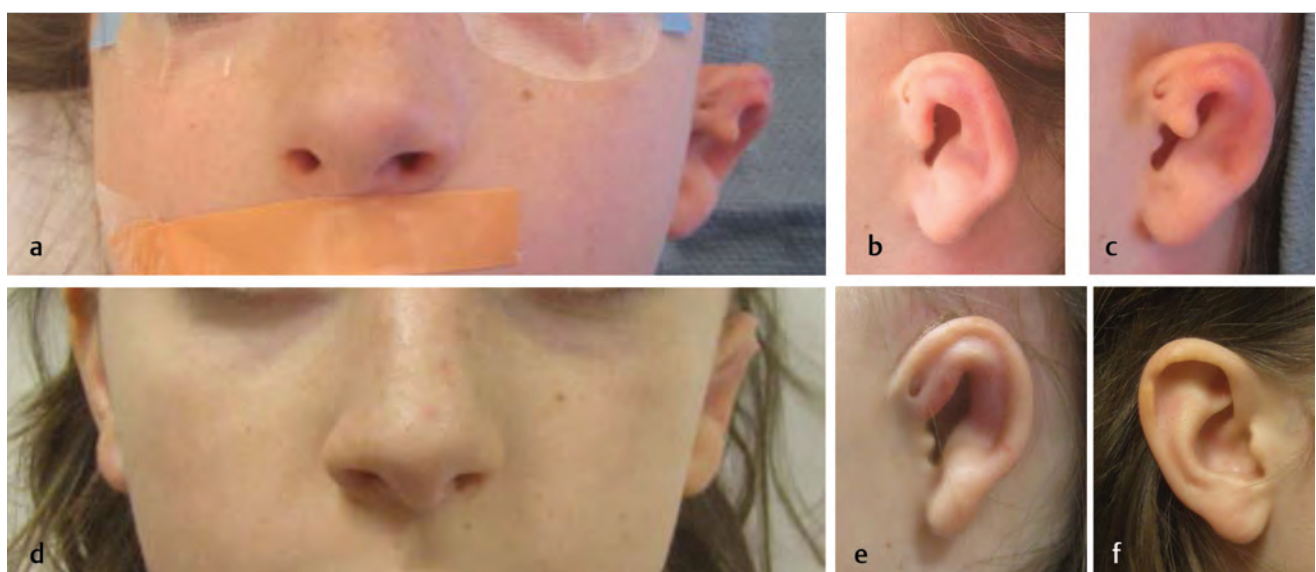


Fig. 20.15 Treatment of a moderate constricted ear. (a-c) Preoperative view shows that the left ear is prominent and vertically short. (d,e) Postoperative appearance after removing the accessory piece of cartilage, setting back the ear, and lengthening the ear using cartilage-mastoid sutures. (f) Contralateral normal ear.



Fig. 20.16 Management of a severe constricted ear. This patient could have been managed by complete ear construction for microtia. However, he had significant scarring in the area because of attempted otoplasty when he was an infant at an outside institution. Because his previous procedure complicated complete ear construction, he was managed more conservatively. (a,b) Preoperative appearance. Note vertically short and prominent ear. (c,d) After setting back and lengthening the ear.

20.5.7 Stahl Deformity

This abnormality describes the presence of a third crus of the antihelix, which can cause the helix to have a pointed appearance. Usually, the deformity is minor and does not cause psychosocial morbidity. Operative intervention may be indicated to treat a severe deformity. The anomaly is improved surgically through an anterior incision at the junction of the helix and scapha, elevating the skin, and resection of the abnormal crus (► Fig. 20.18). If the superior crus of the antihelix is missing, then the excised crus can be grafted or rotated to reconstruct this structure.

20.6 Upper/Middle Third Defects

Congenital anomalies in this area are uncommon and usually consist of abnormally formed structures that can be improved with excisional procedures (► Fig. 20.19). Upper and middle third ear defects most commonly result from trauma. Concha injuries typically do not require cartilage replacement and the overlying soft tissue can be allowed to heal secondarily, closed linearly, or healed with a skin graft. To restore soft tissue over helix injuries with minimal cartilage loss, postauricular skin flaps rather than skin grafts should be used (► Fig. 20.20). If a significant amount of cartilage is absent, then cartilage grafts are

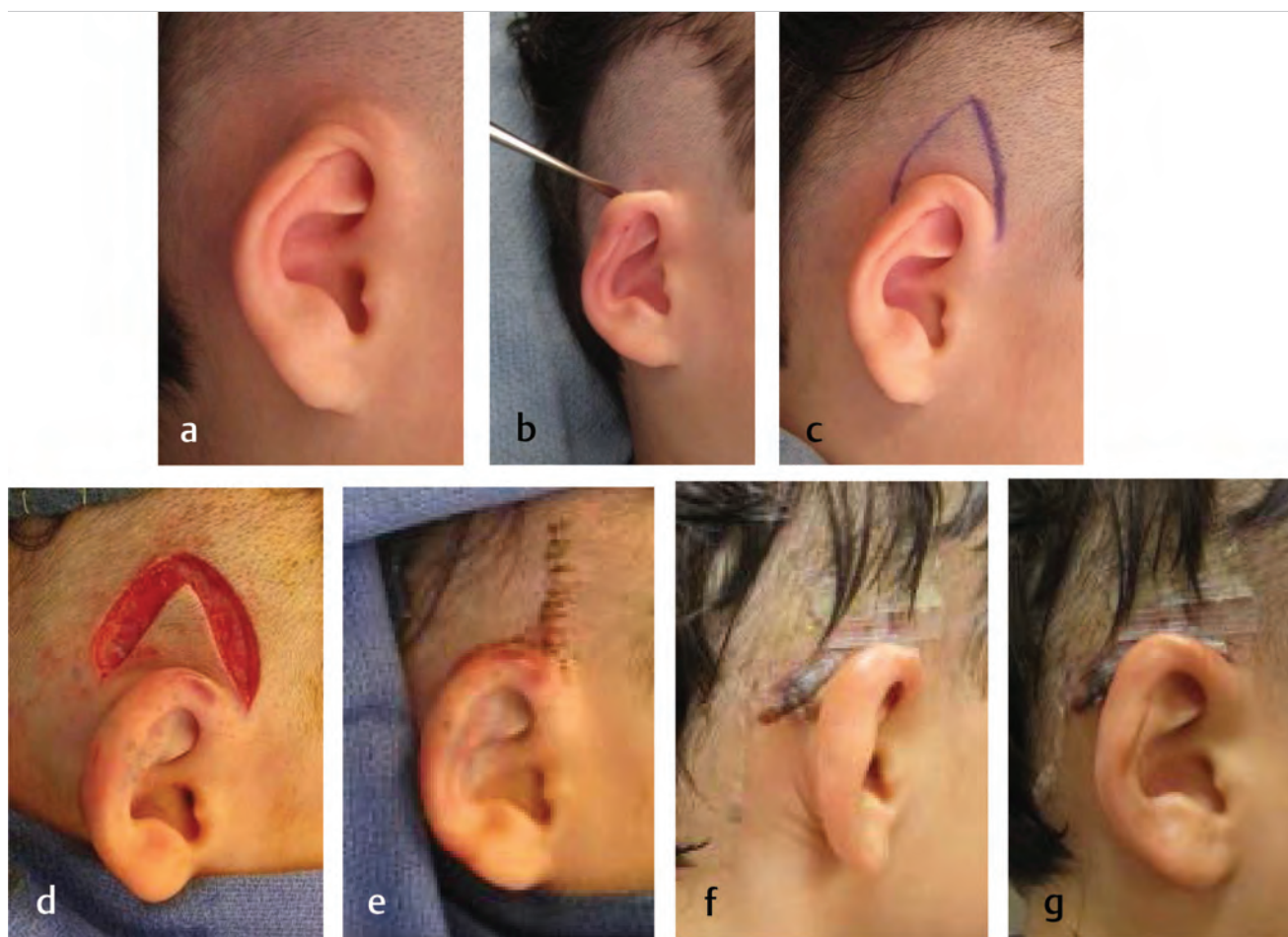


Fig. 20.17 Correction of cryptotia. (a) Preoperative appearance. (b) Illustrating the normal components of the helix by pulling the ear away from the head. (c) Outline of the V-Y advancement flap to recreate the retroauricular sulcus. (d) Flap raised. (e) After advancement of the flap. (f,g) Postoperative views.

placed and covered with postauricular skin flaps in staged procedures. Usually, costal cartilage (hyaline) is preferred over conchal cartilage (elastic) because it is better able to maintain its shape and withstand contractile forces from scarring (► Fig. 20.21).

20.7 Complications

Rarely, a skin abrasion or full-thickness wound can occur with ear molding; molding is discontinued until the area has healed. The most common problem when reconstructing an ear is an unfavorable “cosmetic” outcome. Infection, hematoma, or wound dehiscence is uncommon. Before reconstructing an ear, patients/parents must have reasonable expectations. The ears are tolerant to asymmetry and the primary goal is to improve their appearance on frontal view.

20.8 Conclusion

Ear reconstruction is based on principles. The surgeon must be able to identify the deformity and determine if there are excess

structures, deficient structures, or adequate structures that need to be rearranged. Although many options often exist to improve a specific type of ear deformity, the simplest technique often is preferred.

20.9 Key Points

- Ear molding may correct some anomalies during the first 12 weeks of life.
- Excision of excess structures can be performed in infancy, but other ear procedures are best performed after 3 years of age when most of the ear growth is completed.
- Ear anomalies can be broadly characterized by having excess structures, deficient structures, or adequate structures that need to be rearranged.
- Most ear reconstructions should be done under general anesthesia in the pediatric population.
- Ear reconstruction should aim to give the best symmetry on frontal view.



Fig. 20.18 Correction of a Stahl deformity. (a) Preoperative view. (b) Marking of abnormal crus to be removed and incision along the junction of the helix and scaphoid fossa. (c) Anterior skin flap elevated to expose the abnormal cartilage. (d) Resection of the abnormal crus. (e) Following resection. (f) Repair of cartilage defect. (g) Closure of the incision. (h) Appearance 6 weeks postoperatively.

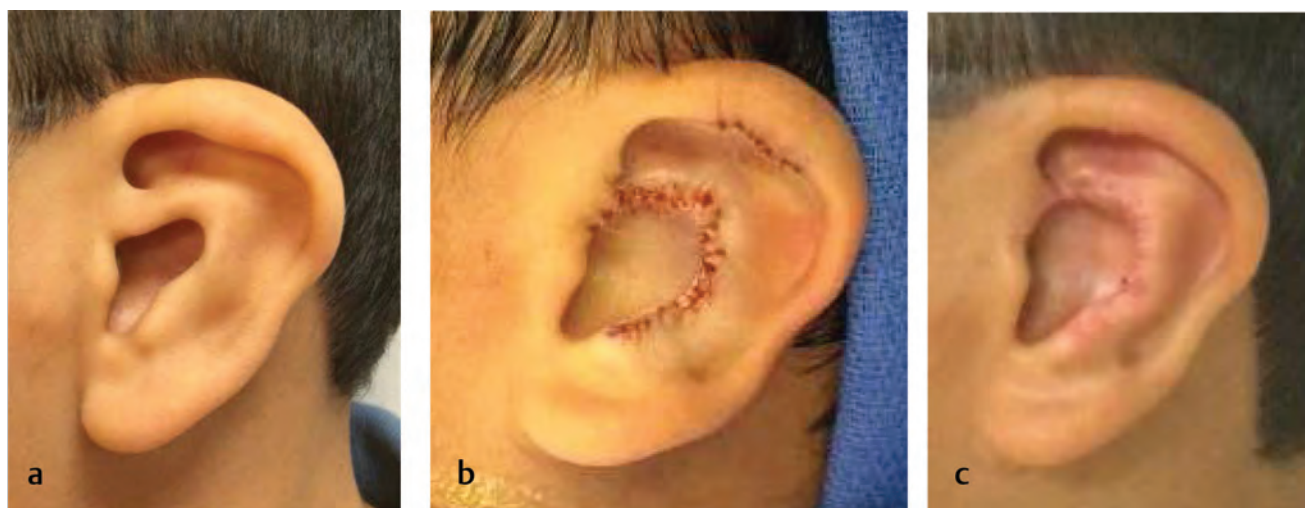


Fig. 20.19 Correction of middle and superior ear anomaly. (a) Preoperative appearance. (b) Excess helical and conchal cartilage was removed. (c) Postoperative result shows opening of concha, improvement of abnormal helical root, and smoother contour of superior helix.

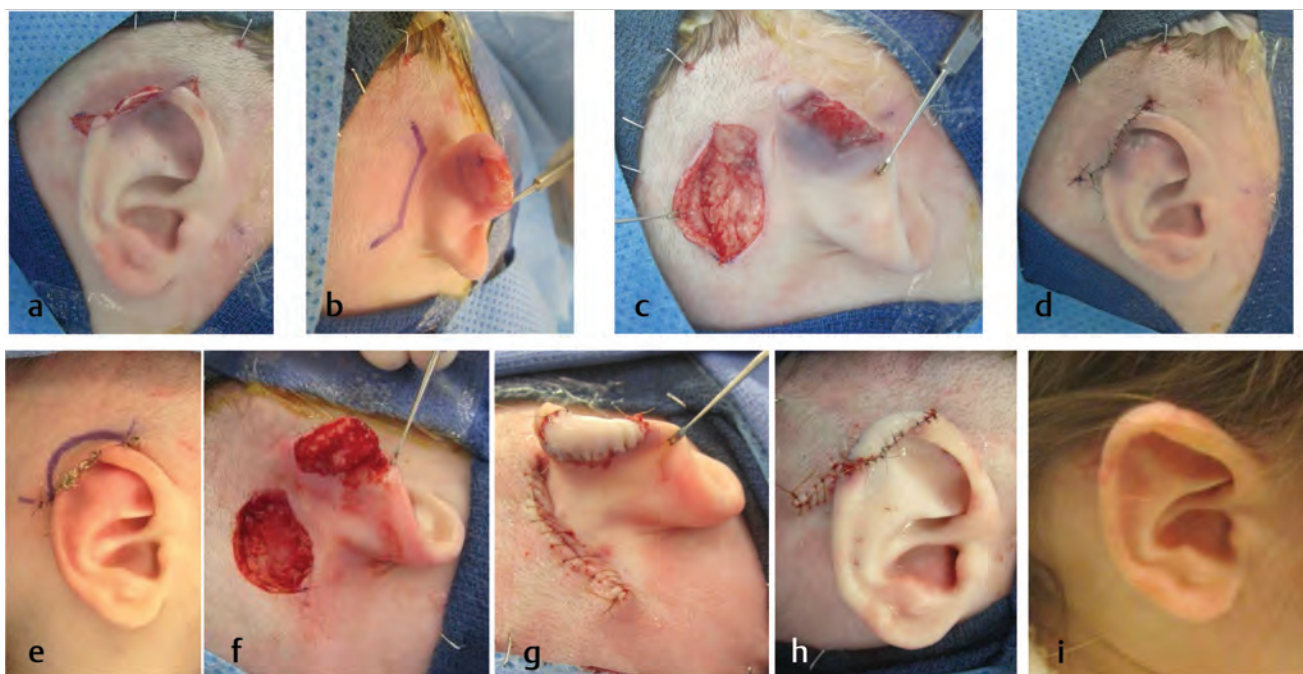


Fig. 20.20 Correction of superior ear deformity following a dog bite injury. (a) Loss of soft tissue and exposed cartilage. (b) Outline of first-stage postauricular flap. (c) Elevation of the flap. (d) The cartilage is placed underneath the postauricular flap. (e) Outline of second-stage division of the base of the flap. (f) Separation of the ear from the postauricular area. (g,h) Flap inset and closure of donor defect. (i) Postoperative appearance.

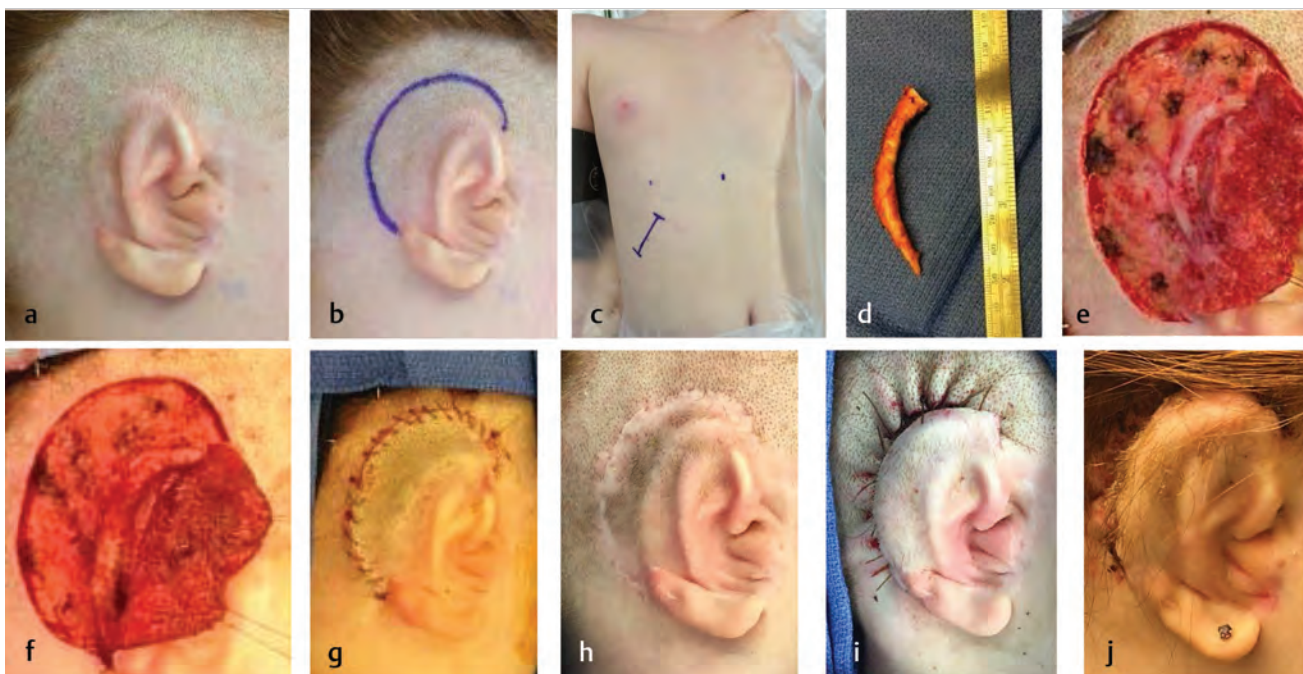


Fig. 20.21 Correction of helix deficiency using a costal cartilage graft. (a) Patient underwent microtia construction by another surgeon. Note deficient helix. (b) Outline of skin flap. (c) Donor site incision. (d) Harvested costal cartilage. (e) Exposed ear construct. (f) Placement of cartilage graft. (g) Closure of skin flap. (h) Prior to second-stage elevation of the construct to improve the definition of the helix. (i) Intraoperative view following separation of the cartilage graft from the mastoid. (j) Appearance 6 weeks postoperatively.

Suggested Readings

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21 Microtia

Akira Yamada

Summary

The auricle is located away from the facial triangle (eye–nose–eye). Therefore, the auricle is not the primary attention of people when they look at someone. The goal of ear reconstruction is to restore/create a natural-looking auricle. The surgeon's focus will be to create the natural helical curve, eliminate unnatural curve, correct excessive protrusion, and restore the missing anatomical parts of the auricle.

Basic knowledge of normal anatomy, proportion, and dimension of the auricle helps prevent mistakes in ear reconstruction. Thin, supple, well-vascularized skin is a must to achieve clear definition of the auricle. It is imperative to create the balance between the amount of skin flap envelope and the cartilage framework to achieve the definition of the auricle.

To be able to create a new auricle from zero, it requires full knowledge and complete skill set of plastic surgery: meticulous skin flap dissection, delicate skin suture techniques, harvesting rib cartilage without perichondrium, elevating temporoparietal fascia flap, and skin harvesting/grafting. Mastering ear framework carving/fabrication techniques requires dedicated, intensive training before starting clinical cases.

Further, it is critical to recognize that what makes a reconstructed auricle beautiful is creating the natural curve of the auricle, especially helix–lobule curve, and concha outline.

Keywords: microtia, secondary ear reconstruction, cartilage framework, hemifacial microsomia, Treacher Collins syndrome

21.1 Clinical Characteristics

Microtia varies from complete absence of auricular tissue (anotia) to a small ear with an ear canal. Nagata classifies microtia into three types: lobule type, small concha type, and concha type. Microtia is nearly twice as frequent in males as in females, and the right-to-left-to-bilateral ratio is roughly 6:3:1. Approximately one-third to one-half of the patients exhibit characteristics of craniofacial microsomia. Brent found that 15% of his 1,000 patients had paresis of the facial nerve. Dellon has shown that the palatal muscles are rarely spared in this syndrome.

21.2 Timing of Surgery

Brent begins ear construction at the age of 6, when the normal ear has grown to within 6 to 7 mm of its full vertical height. Nagata begins auricular construction at the age of 10, and chest circumference has grown over 60 cm at this age, at the xiphoid level. These two different timings could be explained by the difference in the amount of cartilage required.

21.3 Epidemiology and Genetics of Microtia

Microtia (= small ear) is a congenital condition with unknown cause. Prevalence of microtia varies significantly among ethnic

groups (0.83–17.4 per 10,000 births), and is higher in Asian countries for unknown reason. About 80 to 90% of microtia is unilateral, and 10 to 20% is bilateral. There are more than 18 different microtia-associated syndromes with single-gene or chromosomal aberrations; however, there is no causal genetic mutation confirmed to date. Relatively common syndromes associated with microtia are hemifacial microsomia and Treacher Collins syndrome. Isolated microtia rarely runs in families. Treacher Collins Syndrome, inherited in an autosomal-dominant fashion, often presents with bilateral microtia.

21.4 Middle Ear Problem

Ideally, treatments of microtia involve reconstruction of the external ear and the restoration of normal hearing. Hearing impairment in microtia is related to abnormal auditory canal, tympanic membrane, and middle ear. The problem is conduction. Regarding middle ear surgery for hearing restoration, most surgeons presently feel that potential gains from middle ear surgery in unilateral microtia are outweighed by the potential risks and complications for the surgery and that this surgery should be reserved for bilateral cases. The bone-anchored hearing aid (BAHA; Cochlear, Mölnlycke, Sweden; and Ponto, Oticon, Kongeballen, Denmark) has been used since 1977 and does not require a functioning middle ear or patent canal. In microtia patients, BAHA was initially used for bilateral conducting hearing loss. Only 1 BAHA is usually placed because a single aid will stimulate both cochleae simultaneously. Although traditional teaching has been that hearing on a single side is sufficient for speech development and education, evidence indicates audiologic and subjective benefits when treating unilateral conductive hearing loss with a BAHA. Indications are evolving, and reconstructive surgeons and otologists should work together to achieve long-term success in hearing restoration and auricular construction.

21.5 History of Autogenous Ear Reconstruction

Sushruta (6 BC) was probably the first surgeon to perform auricular construction, repairing the ear lobe with cheek flap. Tagliacozzi described repair of ear deformities with retroauricular flaps. Early surgical attention focused mainly on traumatic deformities. However, by the end of the 19th century, surgeons began to address congenital defects, in particular prominent ears (Ely, 1881). The concept of microtia repair began in 1920 when Gillies buried carved costal cartilage under mastoid skin and elevated it with cervical flap. Gillies (1937) also used maternal ear cartilage for more than 30 microtic ears; these were found to have progressive resorption. Peer (1948) diced autogenous rib cartilage and placed it in a Vitallium ear mold beneath the abdomen. After 5 months, he retrieved the banked mold, opened it, and harvested the ear framework for microtia repair. Steffensen (1955) attempted to use preserved cartilage and found it progressively resorbed.

Tanzer is the father of modern auricular construction. Tanzer's excellent results (1959) determined the dominance of autogenous construction with rib cartilage in reconstructive surgery. In the United States, Brent succeeded Tanzer in the early 1970s, and Brent's artistry has influenced reconstructive surgeons even today. Nagata's method (two-stage method) gradually gained popularity, and currently is the most popular choice. To this day, autogenous cartilage remains the most reliable material that produces results with the least complications.

21.6 Alloplastic Implants

Although silicone breast implant survives even today, silicon ear framework is seldom used. Silicon framework, introduced by Cronin in 1966, like any other types of synthetic materials, extrudes, causes infection, and loses definition in the long term. Porous polyethylene (PPE) framework, a newer synthetic material, was first introduced by Reinisch in 1991. The disadvantage of the Medpor framework includes use of temporoparietal fascial flap, long-term risk of implant exposure or loss, and compromise of any future autogenous options.

21.7 Prosthesis

Controversies remain regarding appropriate treatment selection for patients with major ear deformities. This is especially true for severe trauma cases, such as extensive third-degree facial burns. Osseointegrated auricular prosthetic reconstruction is complementary to other approaches and provides a reasonable alternative for poor autogenous options and/or failed autogenous reconstruction. The disadvantages of a prosthesis include intermittent soft-tissue problems, long-term maintenance, requirement of a new prosthetic every 2 to 5 years, ongoing cost, inability for future autogenous options, and the need for a compliant patient.

21.8 Total Auricular Construction in Microtia: Author's Methods

21.8.1 Patient Assessment

About 20 to 60% of children with microtia have associated anomalies or an identifiable syndrome; therefore, individuals with microtia should be examined for other dysmorphic features. Microtia is a common feature of craniofacial microsomia, mandibular dysostoses (e.g., Treacher Collins and Nager syndromes), and Townes-Brocks syndrome. These conditions should be considered among the differential diagnoses when evaluating an individual with microtia.

Facial Symmetry

Asymmetry of the face will make it complicated to locate the ear position. The location of vestige skin is sometimes misleading, and it influences the decision-making of surgeons. Even for expert surgeons, it is tempting to make the auricle based on the current location of the vestige skin, so the surgeons could apply their typical techniques.

Skin Envelope

Assessing the available, soft elastic skin is critical, since it will determine the volume, dimension, and size of the three-dimensional (3D) structure of the ear framework needed. Unbalance between skin envelope and framework may make definition of the auricle poor. Scar interferes the normal stretching of the supple skin envelope and may prevent good definition of the auricle. Scar along the course of the STA (superficial temporal artery) could be a sign of a severed STA, the pedicle of temporo-parietal flap, which is an important salvage tool for auricular reconstruction. This is especially important for microtia patients with craniofacial deformities, such as Treacher Collins syndrome. These patients may have bicoronal scarring due to previous skeletal reconstruction.

Vestige Skin

Since microtia has inherent skin envelope deficiency, how surgeons utilize vestige skin strongly influences the definition of the new auricle. Surgeons need to evaluate the location, shape, and volume of the vestige. These factors will influence the design of skin incision, and surgical strategy. If the vestige skin is located inside or near the auricular rectangle, the vestige skin is ready to be utilized for auricular construction. If the vestige is located far away from auricular rectangle, staged transposition of vestige skin may be necessary prior to the framework placement procedure.

Hairline

Surgeons need to recognize the presence of low hairline at the time of initial evaluation to formulate their surgical strategy (► Fig. 21.1). The extent of a low hairline influences the choice of surgical procedures. If the hairline exceeds beyond the upper one-third of the auricular framework, either preoperative laser

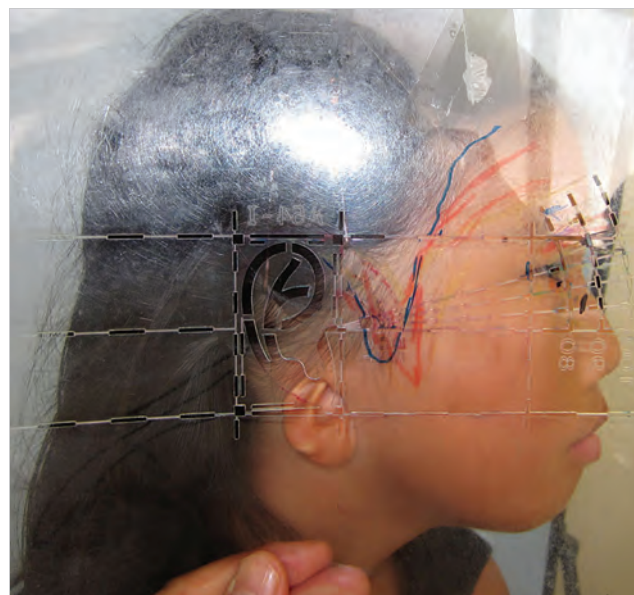


Fig. 21.1 An 8-year-old female with microtia, complicated with low-set vestige plus low hairline, diagnosed with ear positioning template.



Fig. 21.2 The normal auricle is approximately 20 mm posterior to sideburn at the cephalic end of the tragus.

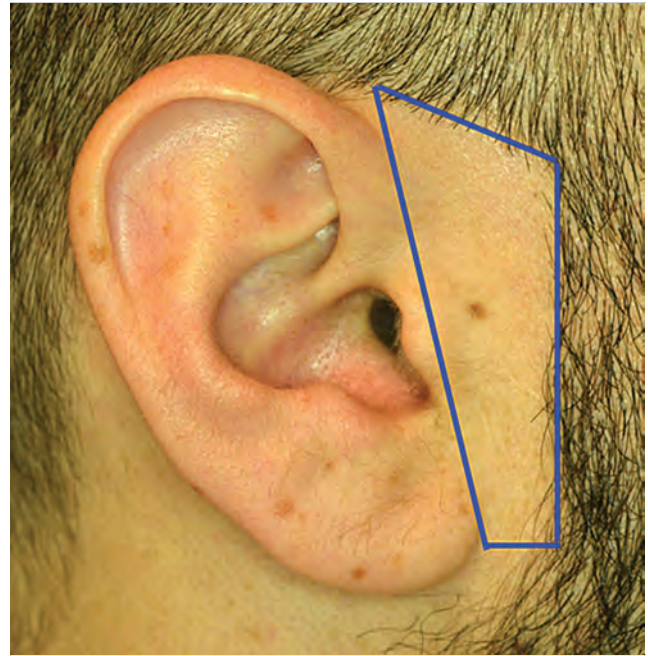


Fig. 21.3 There is a nonhair trapezium-shaped skin area between the normal auricle and sideburn.

hair removal or intraoperative, fascia flap coverage of the framework, after hair-bearing skin removal, may be necessary. If the extent of a low hairline is mild, hairs down to the scapha fossa can be removed during the second-stage procedure (ear elevation).

21.8.2 Trapezium Space behind Sideburns

The sideburn is often missing in microtia patients, especially in hemifacial microsomia. If the patient has a normal sideburn, it is an anatomical landmark useful for locating the proper ear position. Anthropometrical study shows that the auricle is normally located approximately 20 mm behind the sideburn (► Fig. 21.2). There is a trapezium-shaped non-hair-bearing space between the sideburns and auricle normally (► Fig. 21.3). Avoid placing the ear framework in this trapezium to prevent anterior inclination of the auricle.

21.8.3 Location of the Normal Auricle

Leonardo da Vinci analyzed the facial proportion in the same way that anthropologists do today. Recognition of the normal location of the auricle is critically important to avoid misplacement of the construct (► Fig. 21.4). The normal auricle is located at the edge of the face mask (► Fig. 21.5). Surgeons should avoid placing the new auricle inside the face mask, especially in hemifacial microsomia.

21.8.4 Auricular Rectangle

A critical step in preoperative planning is to identify the “auricular rectangle,” where the framework will be placed (► Fig. 21.6).

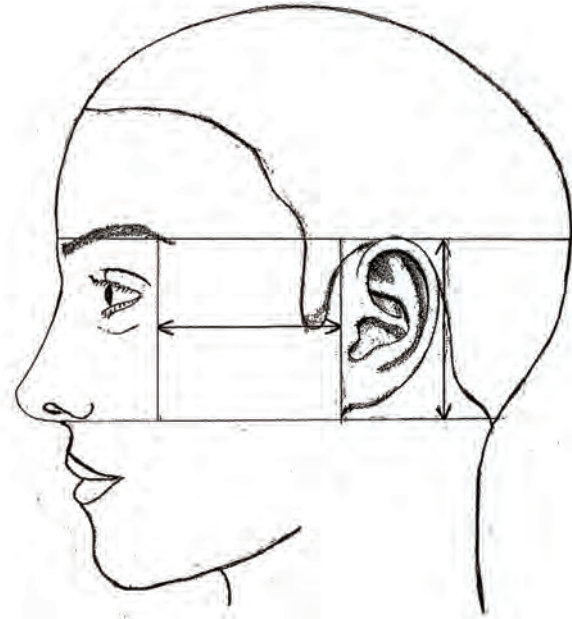


Fig. 21.4 The normal auricle is one ear length behind the lateral orbital rim.

Identifying the auricular rectangle is more challenging in hemifacial microsomia. Once the auricular rectangle is identified, the next step is to evaluate the relationship between the auricular rectangle and vestige skin. A decision has to be made whether vestige skin can be utilized or not.

21.8.5 Ear Positioning Template

An ear positioning template (EPT) is a tool to identify the auricular rectangle, which the author developed with Nagata. It is especially helpful for secondary auricular construction, when many landmarks are missing. Harada and Yamada modified EPT in 2011; more anthropological references are added to the template, and a new template is made of acrylic that facilitates easier surgical marking (► Fig. 21.7). EPT may be used to visualize the extent of low hairline (► Fig. 21.1).

21.8.6 Auricular Curve Analysis

Harada and Yamada developed an auricular shape classification based on a curve ratio analysis study. They identified two key lines in ear shape. The helix-lobule curve and the concha outline curve. There are three major helix-lobule curves (types A, B, and C) and two major concha outline curves, types 1 and 2 (► Fig. 21.8). Normal ear shape may be classified into six groups based on this analysis, and six types of auricular framework

templates were developed as a guide for ear framework construction (► Fig. 21.9).

21.8.7 Auricular Template

When surgeons plan for auricular reconstruction, most use a template as a guide for fabricating the framework. The most widely used method has been to trace the normal auricle with transparent film. The manual tracing tends to be too big and gives poor definition due to skin deficiency. Nagata developed a single ear template for auricular construction. Harada and Yamada then created six types of templates in 2009. The aim of the six templates is to express the subtle, individual difference of the auricular shape rather than a single “ideal” template for all patients.

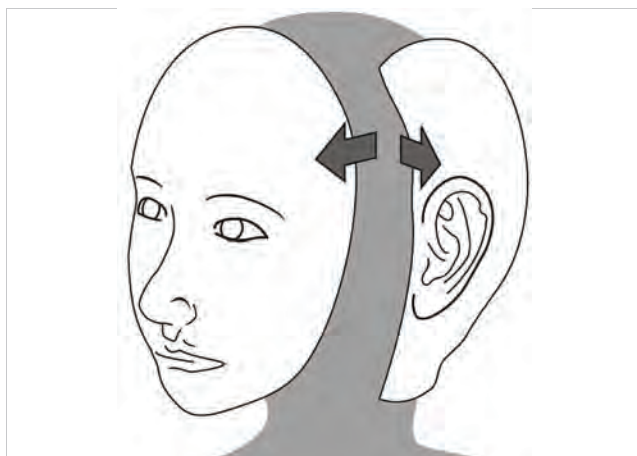


Fig. 21.5 The normal auricle is located immediately behind the face mask.

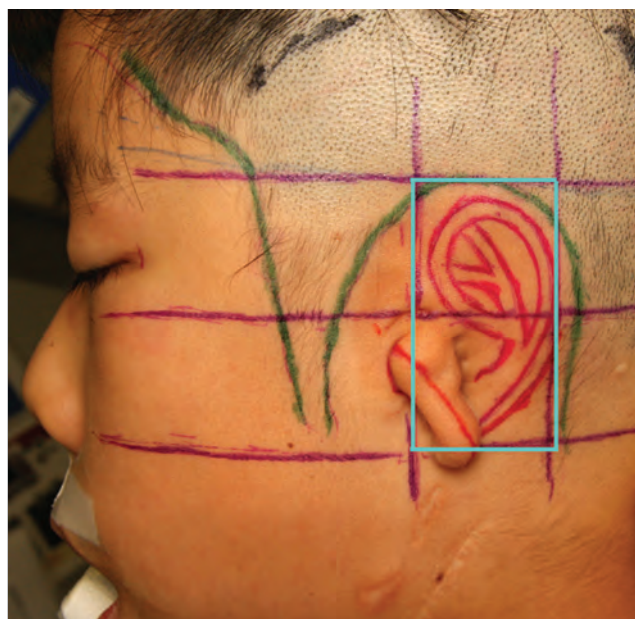


Fig. 21.6 The key to avoid malpositioning of the new auricle is to identify the auricular rectangle.

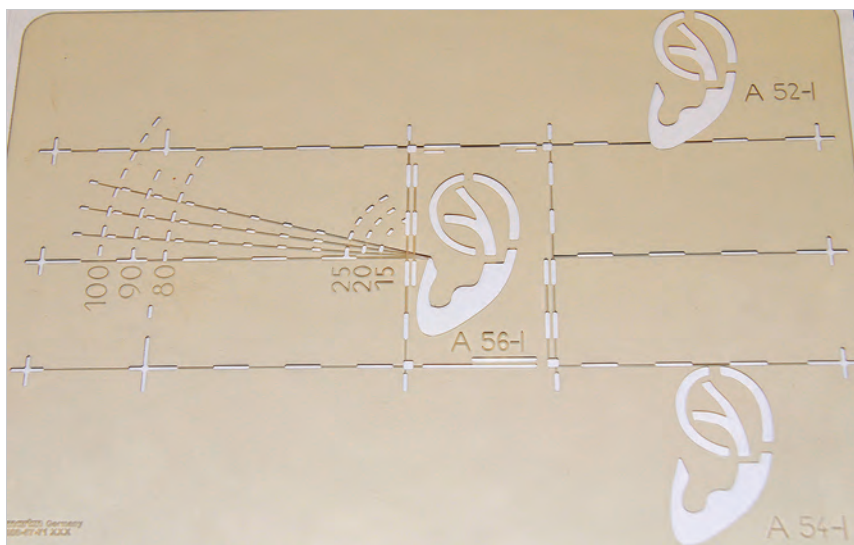


Fig. 21.7 Ear positioning template is a useful tool to identify the proper new auricle location.

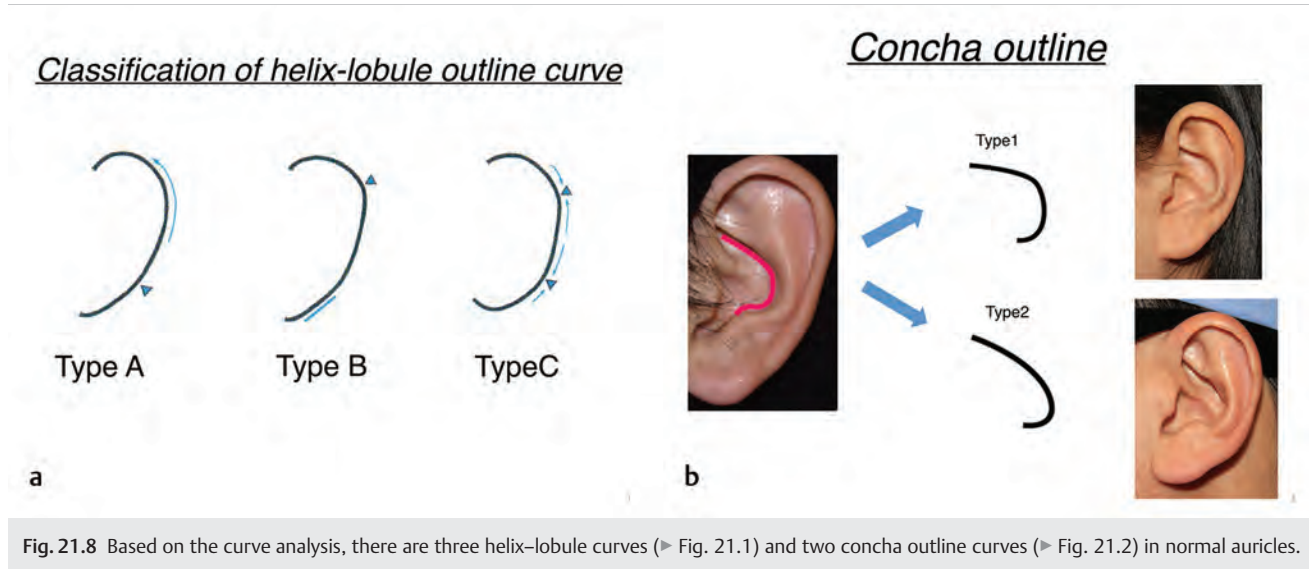


Fig. 21.8 Based on the curve analysis, there are three helix-lobule curves (► Fig. 21.1) and two concha outline curves (► Fig. 21.2) in normal auricles.

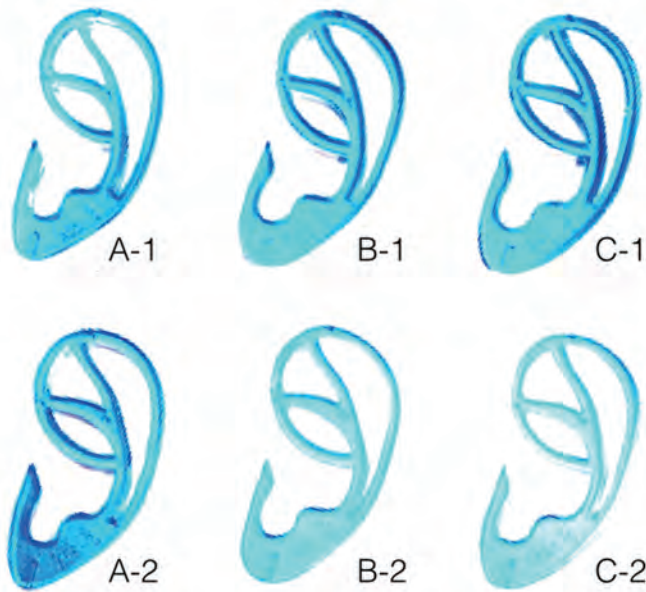


Fig. 21.9 Six types of new auricular template, based on the curve analysis.

21.8.8 First Stage of Total Auricular Construction

Markings

After the induction of general anesthesia, the author performs markings with permanent marker before the surgical preparation. He uses an EPT as a guide for the markings (► Fig. 21.10).

Patient Position

The author uses semilateral position to facilitate simultaneous access to the auricle site and cartilage harvest. Since typical

duration of surgery will be 5 to 7 hours, preventive measures of pressure sore are mandatory. The neck is placed in neutral position to prevent C1 and C2 rotary subluxation.

Skin Flap Preparation

Lobule Splitting Technique for Lobule Type Microtia

Nagata solved the problem of skin shortage in typical lobule type microtia. Creating a deep concha bowl may be achieved by splitting the lobule into two flaps (► Fig. 21.11). Contrary to Tanzer/Brent (full-thickness) lobule rotation, splitting the

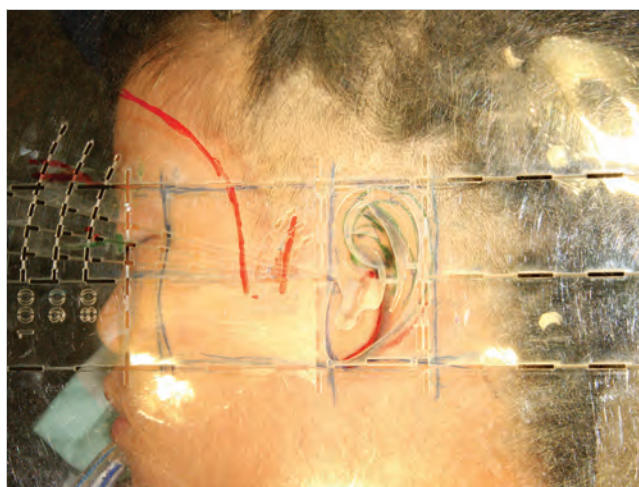


Fig. 21.10 Ear positioning template is applied to identify the auricular rectangle.

lobule creates two flaps: anterior and posterior flaps. Splitting continues down to the fascia level. After completion of the lobule split, the anterior flap is transposed backward to cover the anterior lobule portion of the framework. The posterior flap is moved anteriorly to cover the posterior aspect of tragus and concha. A subcutaneous pedicle is preserved for the posterior skin flap to maintain the vascular supply.

Skin Incision for Small Concha Type Microtia

Nagata defines small concha type by the presence of small indentation in the concha region. The skin incision is made along the margin of small indentation. The indentation is turned inside out to form an inverted cone pocket to cover the framework. The remaining procedure is similar to lobule type microtia.

Skin Incision Design for Concha Type Microtia

Nagata originally (1994) described the skin incision with a posterior V-shaped design (► Fig. 21.12). Recently, Nagata modified the skin incision (► Fig. 21.13) at the posterior aspect of the lobule. In case of concha type with a higher location of vestige than normal, a W-shaped incision has better freedom to transpose the lobule into the optimal location.

Removing Vestige Auricular Cartilage

The author removes all of the remnant auricular cartilage in lobule type microtia. Excising vestige cartilage will create a space to accommodate a new framework. In contrast, in concha type microtia the part of remnant concha cartilage should be preserved as a cuff to facilitate smooth transition of concha cavity.

Skin Pocket Dissection

The author uses small straight blunt scissors, aiming to create 2-mm-thick skin flap. The author does not use epinephrine injection for hemostasis purpose, because injection of fluids makes dissection less accurate, and it may cause vascular

compromise of the delicate skin flaps. The extent of the skin pocket dissection usually goes beyond the hairline border, up to 1 cm beyond the hairline. The author does not violate the trapezium-shaped space in front of the new auricle (► Fig. 21.3) to prevent anterior inclination.

Harvesting Costal Cartilage

Harvesting rib cartilage with perichondrium left at the donor site is a safer technique than harvesting cartilage with perichondrium, in terms of pneumothorax risk. Preserving perichondrium at the donor site, and placing extra diced cartilage into the perichondrial sleeve can regenerate cartilage/bone. This technique may cause less chest wall deformity. If there is clinical suspicion of pneumothorax, a portable chest X-ray, while under general anesthesia, should be taken for possible chest tube placement.

Auricular Framework

Creating an auricular framework is a difficult technique to master. There are several training modules available (► Fig. 21.14), which may be helpful prior to treating patients. The architecture of the auricular framework is critical to create the delicate definition of the auricle. Nagata type 3D framework is distinct from previous 2D framework: the helix connects the bottom of the base frame (first level), climbing as a spiral staircase, to the top of the frame (second level). The tragus is also unique to Nagata type 3D framework and is covered by a posterior W-shaped skin flap, using the lobule split technique.

Bolster Sutures

Nagata uses bolster sutures for his postoperative dressing that was popularized by Tanzer. Brent uses suction drain postoperatively. At the first stage of total auricular construction, the author uses suction only temporally during the surgery to visualize optimal skin adaptation to the framework (► Fig. 21.11.f). After completion of bolster sutures, the author removes the temporary suction. Based on the author's experiences, bolster sutures cause minimal hematoma formation. Dense roll and excessive fixation may be the cause of pressure necrosis underneath a bolster.

21.8.9 Second Stage of Total Auricular Construction (Auricular Elevation)

The difficulty of auricular elevation is often underestimated. The normal auricle is separated from the mastoid area by supporting cartilage. The simplest way to divide the auricle from head is to place skin graft in between head and auricle without skeletal support. Since the resultant defect after division of auricle from head is relatively large, popular choice of skin donor site has been groin area. The disadvantage of auricular division with groin skin grafting include less optimal elevation, persistent edema of the auricle, pubic hair growth, mismatched skin color, and difficult skin cleaning. To overcome the disadvantage described earlier, Nagata proposed more complex auricular elevation than the author routinely performs (► Fig. 21.15).



Fig. 21.11 (a) A 10-year-old child with anteriorly inclined lobule type microtia. (b) Topographic and skin incision markings. (c) Lobule split is completed. (d) Anterior lobule flap is transposed posteriorly, and posterior W-shaped skin flap is transposed anteriorly. (e) The extent of skin pocket dissection is shown. (f) A three-dimensional (3D) framework and 54-mm type A1 template. (g) Immediate postoperative view with temporally suction catheter in place. (h) The postoperative view after the first-stage surgery.

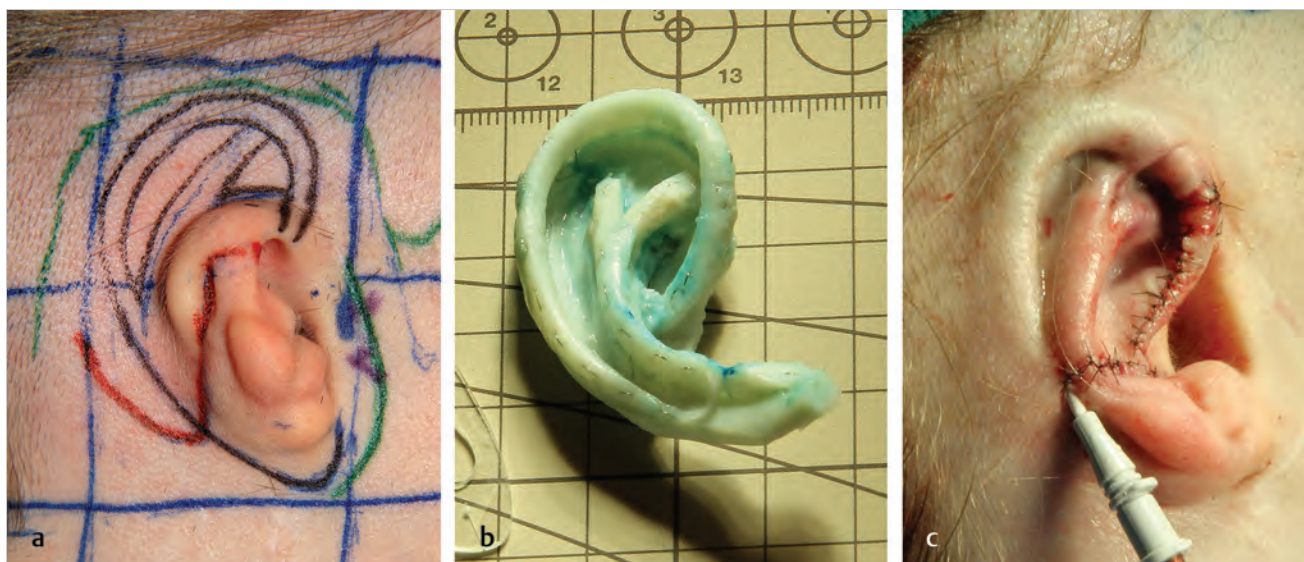


Fig. 21.12 (a) V-shape skin incision for concha type microtia. (b) A three-dimensional (3D) framework without tragus. (c) Immediate postoperative view, with temporal suction in place.

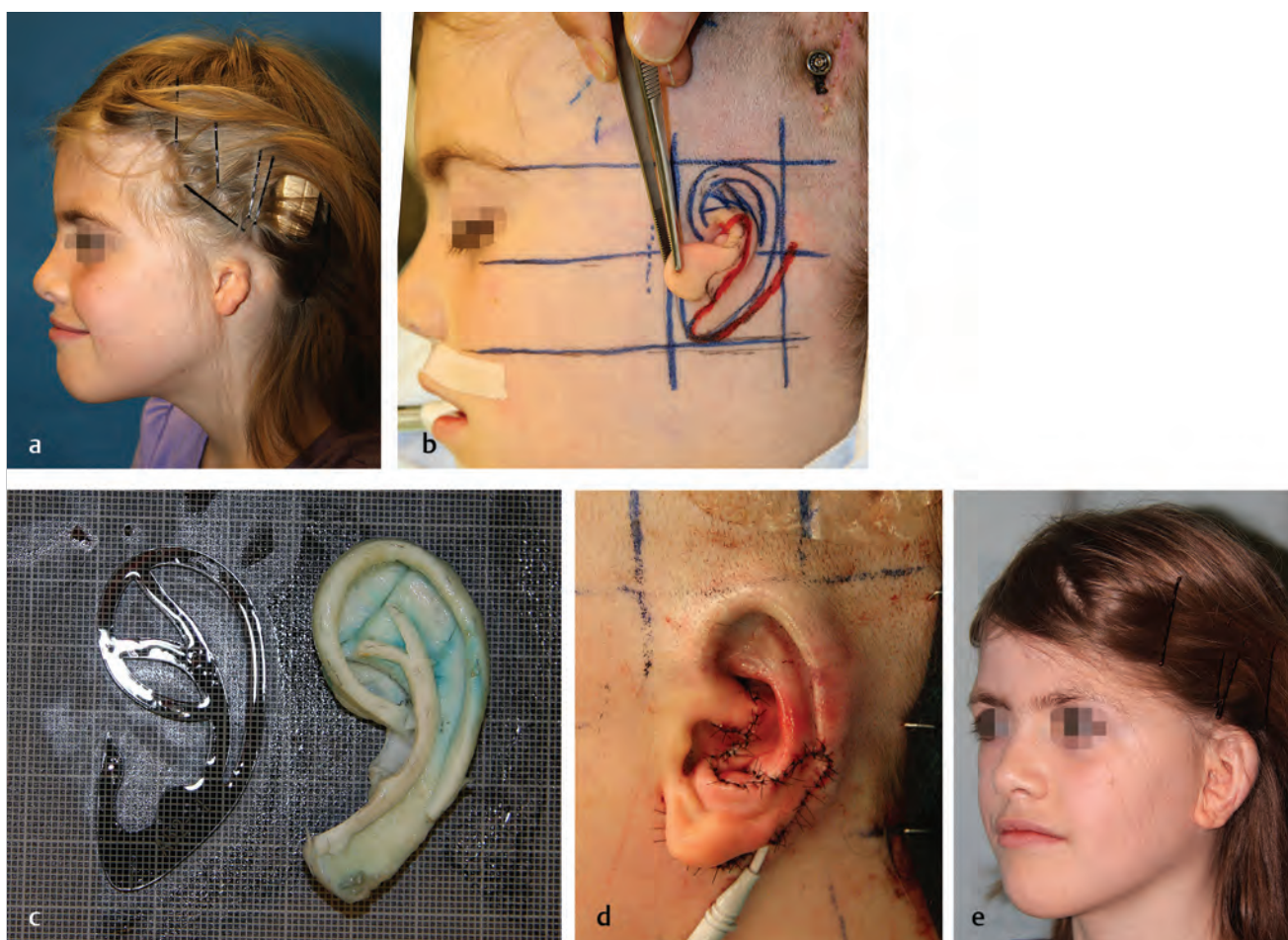


Fig. 21.13 An 8-year-old child with concha type microtia. (a) Preoperative profile. (b) Red marking shows the modification of the skin incision design to bring down the lobule more than 15 mm. (c) A three-dimensional (3D) framework with 52-mm type A1 template. (d) Immediate postoperative view. (e) The postoperative view after ear elevation.



Fig. 21.14 Example of ear carving training, developed by the author. Carrot framework parts and framework made of rib cartilage model are shown.

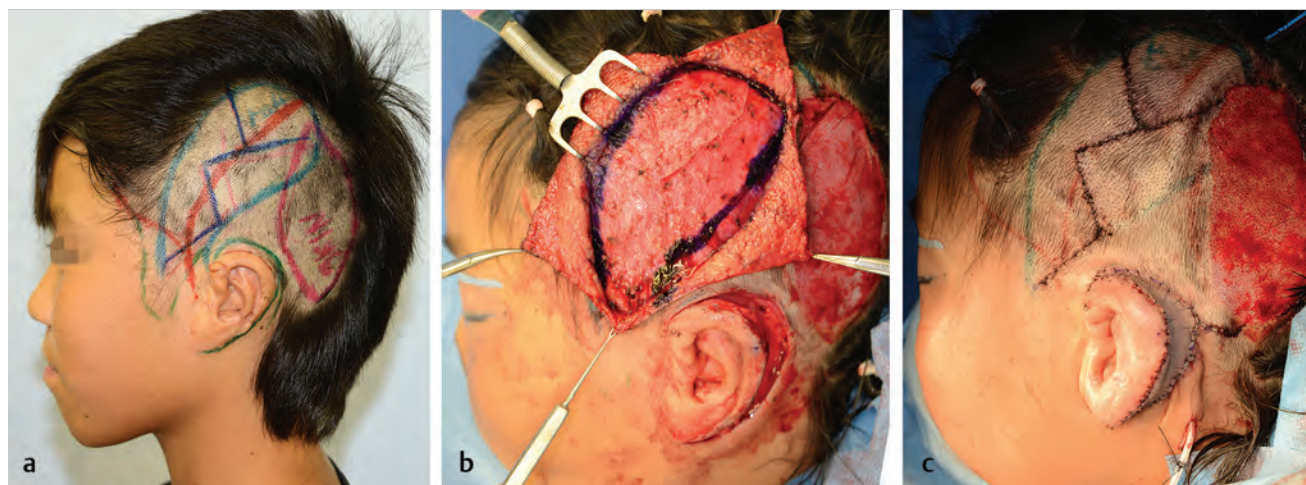


Fig. 21.15 The second stage (ear elevation) for a 10-year-old child with lobule type microtia. (a) Preoperative markings. (b) The marking for harvesting temporoparietal flap. (c) The view immediately after the completion of ear elevation.

Raising Temporoparietal Fascia Flap

The author uses the temporoparietal fascia flap (TPF) to cover the entire posterior aspect of the auricle, not just cover the cartilage block for elevation. If low hairline is mild (hairs cover helix to scapha fossa), TPF enables surgeons to perform intraoperative hair removal. The author routinely uses Doppler ultrasound to plot the course of STA on the temporal skin. If TPF is not available, either deep temporal fascia or a fascia flap based on posterior vasculature is used. If local fascia flap is not available, the third option may be a free vascularized fascia flap. Alopecia has been reported as a complication of the TPF harvest.

Harvesting Scalp Split-Thickness Skin

The author does not use groin skin for auricular elevation because of its poor skin color match and potential pubic hair growth. Nagata proposed the use of split-thickness scalp skin, which has better color match with the auricular skin.

Cartilage Block for Elevation

Many agree that without placing block underneath the ear framework, it is not sufficiently elevated from the head. The author makes an intraoperative impression to create a 3D template to fabricate a cartilage block for sulcus construction (► Fig. 21.16).

21.9 Hemifacial Microsomia

Auricular construction in hemifacial microsomia (HFM) is complex and challenging. Even in mild form of mandibular underdevelopment, vestige skin tends to be located inferior and anterior. A low hairline is present, and patients have a low set ear canal (► Fig. 21.17). If surgeons simply apply the standard formula of auricular construction (place the auricle where vestige is located), the auricle tends to be located in the face mask, which may be aesthetically unacceptable. Surgeons should

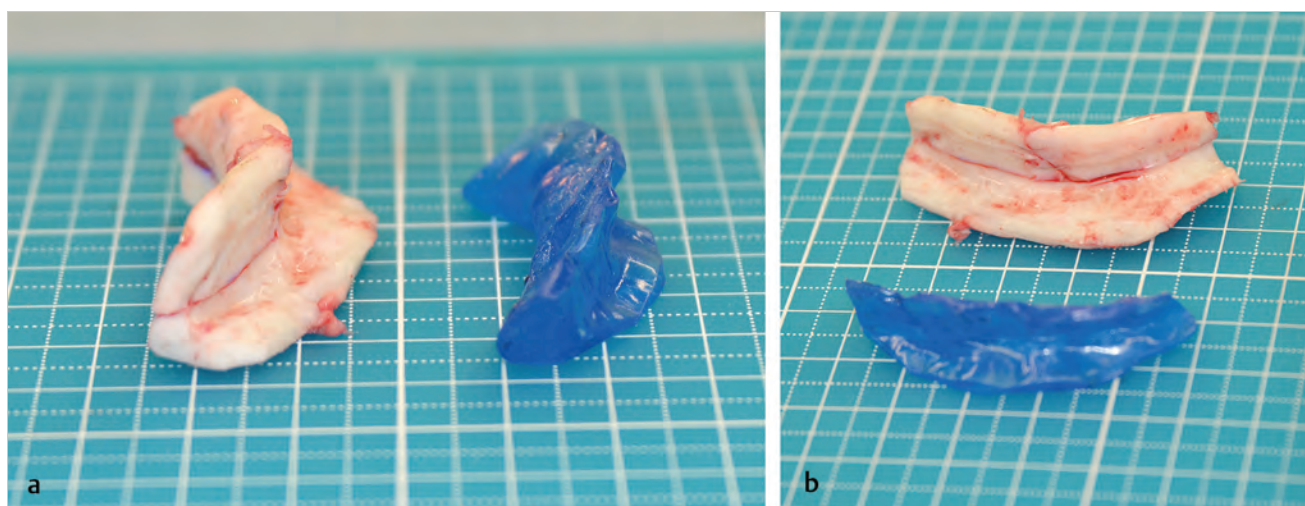


Fig. 21.16 The view of the fabricated cartilage block for ear elevation with thermoelastic intra-operative template for elevation. (a) Caudal side view. (b) The posterior view.



Fig. 21.17 An 8-year-old with low-set, low hairline microtia.

avoid placing the new auricle inside the face mask. It is a common mistake to place the new auricle in an anteroinferior location, trying to avoid the low hairline. In HFM, skeletal correction prior to ear construction may help to place the auricle in proper location. It is notable that laser hair removal works best for black hairs with multiple sessions (could be more than 10 sessions to complete), but currently it is not effective against blond hairs. The author has found that in the microtia patient with severe HFM there is a higher risk of cartilage resorption.

21.10 Treacher Collins Syndrome

The patient with Treacher Collins syndrome has distinctive facial features: high nose, deficient lateral orbit/zygoma, small chin, and smaller overall facial size. Placing a large auricle may be disproportional and unnatural. The size of the new auricle, therefore, needs to be smaller than average; the author usually selects the auricle size 45 to 50 mm in length to balance with the rest of the face. Microtia in Treacher Collins syndrome may accompany a low hairline. To eliminate hairs from the auricle, laser hair removal is one option; the other option is applying a local fascia flap (e.g., temporoparietal fascia or fascia flap based on posterior vascular pedicle) to cover the upper one-third of the framework.

21.11 Unsatisfactory Outcome

The cause of unsatisfactory outcomes are multifactorial: improper (1) evaluation of the deformities, (2) selection of the surgical procedures, (3) recognition of the facial asymmetry, (4) assessment of low hairline, (5) selection of the new auricular location, (6) inclination of the auricle, (7) timing of the surgery, (8) amount of costal cartilage, (9) cartilage harvesting techniques, (10) skin pocket dissection, (11) lobule spiriting techniques, (12) shape, volume, size, and width of the framework, and (13) postoperative care.

21.12 Complications

The overall complication incidence in ear reconstruction is reported to be 16.2% on average with a range of 0 to 72.9%. Probably the most serious complication in total auricular construction is cartilage infection, leading to the entire extrusion of the ear framework. To prevent this disaster, even small skin necrosis has to be addressed immediately. Skin necrosis along the helical rim may be repaired with local skin



Fig. 21.18 A 16-year-old female with lobule type microtia. Tissue expander reconstruction was performed at the other institution. (a) Preoperative view before secondary construction. (b) The 56-mm type A1 template is applied to evaluate the shape. (c) A three-dimensional (3D) framework. (d) The upper two-thirds of the new framework is covered with temporoparietal flap. The lower one-third of the framework is covered with local skin flap. (e) Split-thickness scalp skin was grafted over the fascia. (f) The postoperative view of the patient.

flap, but skin necrosis in medial portion, such as concha bowl, usually requires local fascia flap to salvage it. Long-term complication includes collapse of ear framework and extrusion of wires. Total auricular construction in HFM has a higher frequency of complications, such as malposition and resorption of the auricle.

21.13 Secondary Construction

Secondary construction is challenging, but it is not impossible. In many unsatisfactory outcomes, patients have lost supple skin, have scars, and have resorbed frameworks. To create a smooth, natural-looking auricle in such situations, surgeons should take an aggressive approach: discard all scar tissues, deformed framework, and damaged skin envelope, and then replace them with well-vascularized supple, thin skin envelope, and well-planned framework. In secondary cases, temporoparietal fascia flap is a powerful workhorse to provide a thin envelope to cover the framework (► Fig. 21.18). If the local fascia flap is not available, distant free fascia flap is an option. Brent described the use of contralateral TPF plus hair-bearing skin to restore temporal hairline plus auricular construct.

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22 Nasal Reconstruction

Alexander Facque and Peter J. Taub

Summary

While minor, and frequently nonoperative, injuries to the pediatric nose are common, major injuries are relatively rare. Acquired defects can occur as early as infancy due to pressure-related necrosis of the septum, columella, or ala during nasal oxygen administration as in continuous positive airway pressure (CPAP). The more common cause of nasal defects occur during childhood as a result of animal bites and burns. All such injuries are important from a psychological and developmental standpoint. The importance of self-image plays into the psychosocial development of the pediatric patient, and reconstructive efforts should be made in some attempt before the child starts school, approximately by 6 years of age. Defects of the pediatric nose are especially challenging due to the complex anatomy of the nasal structures and the need to grow with the rest of the face. The relative presence or absence of the various nasal components (external skin, cartilaginous or bony support, or internal lining) should be noted and addressed as appropriate.

Keywords: pediatric, nose, ala, dorsum, cartilage, lining, bilobe, paramedian

22.1 Introduction

Given the causes for major pediatric nasal reconstruction are relatively rare, the importance of the nose, from both a psychological and developmental perspective, mandates that they be done with great care. As evidenced by the existence of punitive rhinotomy, described in historical documents since times even before the Code of Hammurabi, undeniably the nose carries a psychosocial importance that supersedes almost any other aspect of the human body. Additionally, from a developmental perspective, the nose is considered one of the growth centers of the face, responsible for some of the development of the midface during childhood as well. Subsequently, defects of the pediatric nose from both congenital and acquired etiologies take on a heightened level of complexity, not only due to the complex anatomy of the nose, but also due to the overall impact that nasal development will have on both psychosocial development and the ultimate development of the face. Furthermore, reconstruction of the pediatric nose carries with it special concerns including whether the reconstruction will grow with the face, how the reconstruction could alter remaining facial development, and if or how future donor sites should be preserved should a future revision be necessary.

22.2 Embryology

At minimum, a cursory understanding of the embryologic development of the nose and face is important when considering reconstruction of nasal anomalies, given these malformations are by definition the result of aberrant embryogenesis and an ability to assess what nasal components or subunits are absent or

malformed will subsequently lead to an understanding as to what will need to be replaced during the course of reconstruction.

As an overview, the pediatric face begins to take shape during weeks 5 to 10 with the formation, fusion, and differentiation of five mesenchymal swellings or prominences: two mandibular, two maxillary, and the unpaired frontonasal. The latter will ultimately be responsible for forming the face above the external nares, including the forehead, interorbital region, and the majority of the external nasal structures, while the maxillary prominences will comprise the oral opening to the external nares and the mandibular prominences will fuse to create structures inferior to the oral opening (► Fig. 22.1).

Initially, development of the nose begins with formation of the nasal placode from localized thickening in the superficial ectoderm of the inferior portion of the frontonasal prominence, immediately above the nasolacrimal groove between the frontonasal and the maxillary prominences. Sensory neurons in the placode differentiate to form olfactory neurons, while thickening of the mesenchyme at the periphery of the placode elevates the ectoderm above the placode to produce nasal pits at week 5. The outer areas of these pits will form the medial and lateral nasal prominences, from the inferior portion of the frontonasal process. The medial nasal prominences ultimately fuse together to form the median nasal prominence at week 8 (leading to the creation of the nasal dorsum) and will also fuse inferiorly with the lateralized maxillary prominences to form the central and lateral portions of the upper jaw, including the philtrum, nasal septum, and the primary palate. The lateral nasal prominences fuse with the more cephalad aspect of the maxillary prominences to form the nasal alae as well as lateral portions of the nose. The maxillary prominences also will go on to form the secondary palate, an example of the interconnectedness of nasal and palatal formation explaining why cleft palate malformations are inherently linked to nasal deformations as well.

22.3 Development

After birth, the nose continues to grow and change with the facial profile throughout adolescence. The infant nose is noted to have less frontal projection with a shorter dorsum, larger nasolabial angle, and rounder nares. The main support of the infant nose is the dorsal septal and upper lateral cartilages, which are considered an extension of anterior cranial base. This cartilaginous septum is considered to be a part of two nasal growth centers, the sphenospinal zone and the sphenodorsal zone, both of which affect the growth of not only the nose itself, but also the surrounding midface. The sphenodorsal zone is described as being responsible for growth in the length and height of the dorsum, while the sphenospinal zone is responsible for sagittal growth, as well as outgrowth of the maxilla. Because of the presence of these growth zones, loss of septal cartilage in infancy and childhood can lead to various acquired

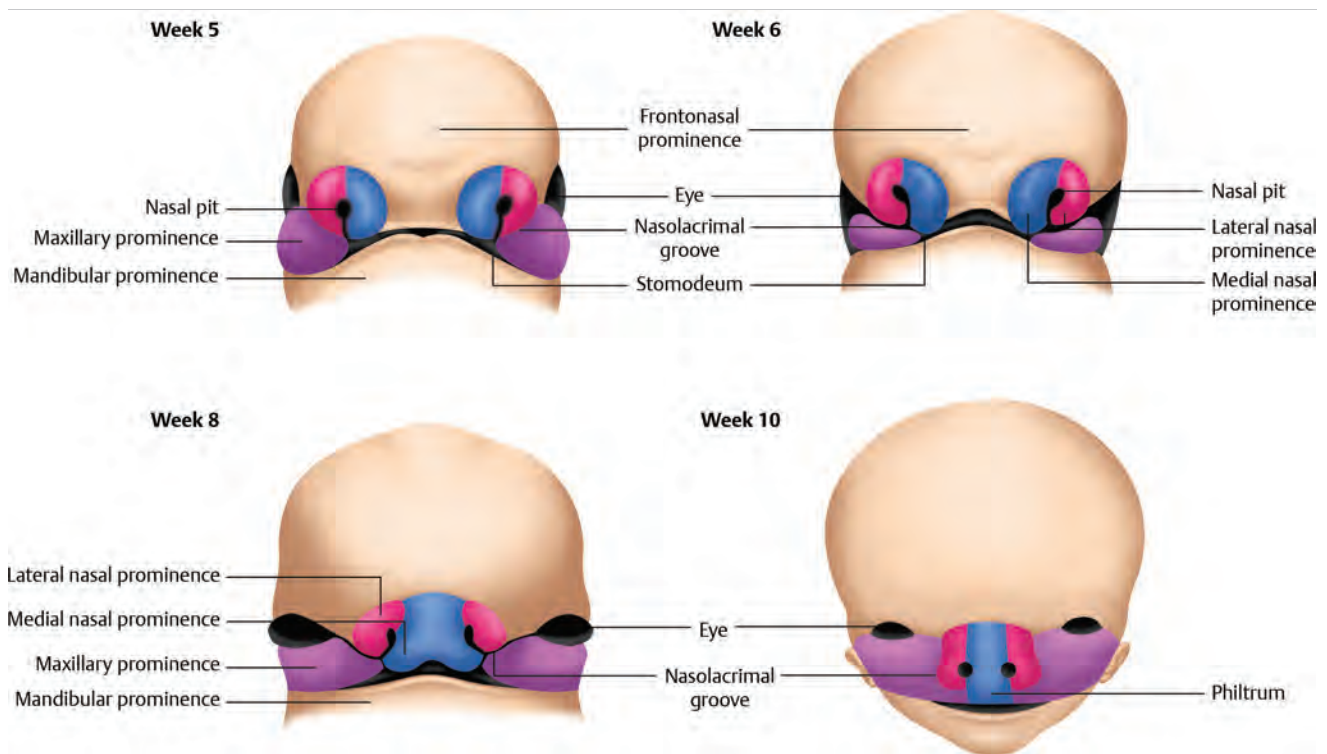


Fig. 22.1 Embryologic development of the nose.



Fig. 22.2 Nasal hemangioma.

facial syndromes involving not only the nose, but also the maxilla and the orbit. Furthermore, surgery on the nose that disrupts these growth centers may have similar and predictable deleterious effects. Nasal growth appears to continue until early adulthood with specific windows of accelerated growth. The two most significant nasal growth spurts occur in the first 2

years of life and during puberty. The end of the period of growth was found to be at 12 to 16 years in girls and 15 to 18 years in boys.

22.4 Diagnosis

Defects of the external framework of the nose are generally obvious on inspection. They can arise from congenital and acquired causes. Congenital defects occur generally as a result of failed or incomplete fusion of the mesenchymal prominences discussed previously. For example, failed development of the frontonasal prominence results in the characteristic deformities seen in frontonasal dysplasia. Another class of congenital defects arises as the result of congenital growths such as benign hemangiomas (► Fig. 22.2), dermoid cysts, and nasal encephaloceles (► Fig. 22.3). A classification scheme developed by Losee et al separated nasal malformations into four broad classes. Type 1, comprising 62%, includes defects arising from hypoplasia or atrophy and agenesis syndromes ranging from whole arhinia to individual subunits including nostril stenosis (► Fig. 22.4), nasal hypoplasia, choanal atresia, and craniofacial syndromes. Type 2 defects (1%) are the result of hyperplasia and duplication exemplified by proboscis lateralis. Atypical clefts comprise type 3 and arise from failure of fusion of medial or lateral nasal prominences and the surrounding maxillary prominence and include the Tessier 0, 1, 2, and 3 clefts as well as their cranial extensions 11, 12, 13, and 14 (► Fig. 22.5). Neoplasm and vascular anomalies are included in type 4 nasal defects (20%). Examples of benign lesions include hairy nevus, glioma, pilomatixoma, neurofibroma, and nasal dermoid (12.4%), as well as benign vascular lesions such as hemangiomas.

Acquired defects of the nose can occur throughout infancy and childhood and include both iatrogenic and hospital-acquired conditions, such as pressure-related necrosis of the septum, columella, or ala during nasal oxygen administration as in CPAP (continuous positive airway pressure). The most common cause of nasal defects during childhood is traumatic, resulting from animal (generally dog) bites and burns.



Fig. 22.3 Nasal encephalocele.

Further diagnostic maneuvers may include one or more imaging modalities to better elucidate the nature of the deformity. If there is a question of depth or involvement of surrounding tissues, a computed tomography (CT) scan or magnetic resonance imaging (MRI) may be indicated. CT scanning will highlight the surrounding bony structures and determine if there is potential involvement of deeper structures. MRI will better elucidate the soft tissues in and around the nose.

22.5 Nonoperative Repair

There are scant nonoperative treatment options for nasal deformities. Scars can be managed similar to other areas of the body, including massage, topical agents, laser therapy, and dermabrasion. Complete loss of the nose may be addressed with a prosthetic device. For the pediatric patient, this, however, may be difficult to maintain with a more active lifestyle and over a long period of time.

22.6 Operative Treatment

22.6.1 Timing of Repair

Due to the importance of self-image in psychosocial development, reconstructive efforts should be made in some attempt before the child starts school, approximately by 6 years of age. As nasal development continues throughout adolescence, waiting until after this was complete would subject the child to possible years of alienation and ridicule although a more definitive repair could be offered at that time. In light of these concepts, best results involve a dual reconstructive approach with an attempt as early as possible (preferably before school age) with knowledge that a subsequent definitive repair may be needed in adulthood for optimal results. Part of this rationale necessarily includes consideration that optimal donor sites for a later, definitive repair may need to be spared during the pediatric, initial reconstructive attempt.



Fig. 22.4 Stenosis of the left external nasal valve.

22.6.2 Nasal Subunit Theory of Reconstruction

In the adult, the nasal defect and their subsequent reconstruction can be analyzed by dividing the nose into aesthetic/recon-



Fig. 22.5 Bilateral Tessier 3 cleft.

structive subunits including the dorsum, tip, alae, sidewalls, and soft triangles. The authors argue that replacing an entire subunit when it is involved in the defect improves the aesthetic outcome of the reconstruction by placing the scars into more camouflaged locations and allowing the reconstruction to follow the pre-existing concavities and convexities of the native nose. Others argue in favor of preservation of as much native tissue as possible. In the pediatric nose, combining subunits, such as the tip and columella and the dorsum and sidewalls, has been suggested to create a simplified approach with just three subunits (dorsum, tip, ala) because in the pediatric nose these borders are often indistinct (► Fig. 22.6). Menick argues that while the borders are indistinct, they are present, and that following the subunit reconstruction principle with these simplifications may lead to over-resection, which is not without its own set of complications. Paramount to discussions regarding reconstruction of the pediatric nose is that a reconstruction must be durable, aesthetically pleasing, and importantly it must be known if the reconstruction will grow with the developing face. Necessarily, if a reconstructive effort is completed that will require further revisions in the adult face, consideration must be placed into choosing tissue that will not preclude subsequent attempts at reconstruction.

Reconstruction Techniques

When addressing nasal defects, assessment begins with an examination of the characteristics of the defect. The location (upper, middle, or lower nose) and the subunit or subunits involved should be noted. Furthermore, the relative presence or absence of the various nasal components (external skin, cartilaginous or bony support, or internal lining) should be noted with a mindfulness of the classic dictum to “replace like with like.” This is important in preventing collapse and further disfigurement. Consideration should be made not only in re-creating the subunits, but also in reconstructing the volume/shape and function of the nose via reconstructing the lining, support, and overlying skin of the nose.



Fig. 22.6 Indistinct borders to the nasal subunits in the pediatric nose.

22.6.3 Nasal Lining

Lacking nasal lining can be addressed using native septal mucoperichondrium, previously scarred skin that may otherwise be discarded depending on the etiology of the nasal defect, skin graft, and/or a variety of turnover flaps. Close attention should be paid to the amount of septal manipulation given the concern for growth arrest discussed earlier. More distant options for reconstructing the nasal lining have been described involving prefabricated galeal flaps, mucosal flaps from the buccal sulcus, as well as islandized frontalis muscle with skin graft. Free tissue can also be folded on itself to create an external surface as well as an internal lining. In this case, one or more secondary debulking procedures will likely be required.

22.6.4 Structure

In defects involving the framework of the nose, cartilage may be harvested from the ear or rib, typically the sixth or the seventh. Again, septal cartilage is typically avoided depending on the age at which the repair is completed due to future growth concerns as well as volume able to be recovered. In older children lacking support to the lower portion of the nose, an L-shaped septal advancement by Millard has been described with good result. A temporo-retroauricular flap as described by Washio has also been described to have the capacity to take bring auricular cartilage to lateral nasal defects, which may be especially useful in alar reconstruction. Paramount to any structural reconstructions involving the nose is the presence of a stable base on which the reconstructions will be placed. The support given by the maxilla should be assessed and supplemented as needed, as is the case in the nasal component of cleft lip/palate repairs, in which bone grafting from the iliac crest to the alveolar gap is performed prior to performing the reconstructive rhinoplasty. Bone for this purpose may be taken from rib, iliac crest, or calvarium depending on the needs and wishes of the patient and/or parent. In older children, in which the concern for growth arrest after manipulation of the septal cartilage is considered negligible, the full array of septal cartilage grafts (spreader, lateral crural, columellar strut) are available to the reconstructive surgeon and can be used in combination with paramedian forehead flaps or other local advancement flaps in a prefabricated or composite fashion.

22.6.5 External Skin

As with elsewhere on the body, skin coverage options cover the spectrum of the reconstructive ladder, from skin grafts (full-thickness preauricular/postauricular, supraclavicular) to local flaps (bilobed, dorsal nasal, cheek advancement, nasolabial, paramedian forehead) to microvascular free tissue transfer. Composite grafts as was discussed in regard to recreating the structural framework of the nose can also be created using any of these methods. With the loss of multiple subunits, the paramedian forehead flap has been described to have the ability to cover extensive skin loss, with acceptable donor site morbidity. Burget describes a three-stage procedure with the caveat that in some patients, preliminary surgeries may need to be performed to debride scar or release contractures that may inhibit the surgeon from gaining an adequate assessment of the

reconstructive needs of the patient. In his description, the first stage of a full-thickness reconstruction provides nasal lining, cartilaginous framework, and coverage with a paramedian forehead flap. The second stage involves elevation and thinning of the forehead flap to better match the thinness of the native nasal skin. And finally, in the third stage, the pedicle is separated and any revisions to the donor site are performed.

22.7 Complications

The major potential complication of not repairing a nasal defect is the social burden, which the child (and his parents) must endure. Infants and younger children may be spared from questions and teasing from other children, but older patients are vulnerable to negative interactions with their peers. Associated nasal compromise from vestibular obstruction or collapse may compromise nasal function. The potential complications following surgical reconstruction include those common to most operative procedures, including bleeding, infection, wound healing problems, persistence of the deformity, and worsening of the deformity. If tissue is recruiting from additional sources, problems may arise that relate to not only the recipient site, but also the donor area.

22.8 Conclusion

Pediatric nasal reconstruction is a complex and interesting topic, with special consideration given to the anatomic and psychosocial importance placed on the developing nose. Special considerations include deciding on the proper timing of the repair, with the primary objective to balance the competing goals of minimizing the traumatic effects on self-image that having a nasal defect may have on the child with the ability to provide a definitive repair. Nasal reconstruction is further complicated by the complex anatomy of the nose, with multiple different types of tissue that will need to be replaced in order to recreate a functional and aesthetically pleasing nose. Once these assessments are complete, nasal reconstruction provides a unique challenge to the reconstructive surgeon, with the potential for very gratifying results to all involved.

22.9 Key Points

- Deformities of the nose may have significant psychosocial implications.
- Reconstruction of nasal deformities must take into account potential future growth of the nose.
- Reconstruction of nasal deformities must take into account specific structures that are involved, including lining, support, and coverage.
- The concept of the reconstructive ladder applies to nasal reconstruction; the concept of the subunit principle may or may not.

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23 Facial Paralysis

Greg Borschel

Summary

The management of pediatric facial palsy begins with the accurate multidisciplinary assessment of the deficit and acute problems, the cause and associated problems, the likelihood of spontaneous recovery and the psychological and social impact on the child and their family. After the management of acute issues (which are uncommon), once there is a decision to operate, an assessment is made of the surgical options. Which one is used depends on the exact deficit, but in general in the pediatric group the problem is the inability to smile. Our preferred option is a two-stage procedure in which the first stage is a cross-face nerve graft and the second stage a free gracilis transfer. This is gone through in detail with the family, and the decision to proceed is made together with them. The outcomes are generally good, with the vast majority of patients who have received the two-stage procedure achieving improved facial symmetry, developing an ability to smile spontaneously, and also very satisfactory results for those receiving free gracilis transfers innervated by the motor nerve to masseter and the complication and revision rates are relatively low.

In terms of the future, recent evidence from the rat model suggests that “super-charging” the nerve graft with a sensory nerve at the time of grafting can improve nerve regeneration. Brief periods of electrical stimulation of a proximal nerve stump can also enhance nerve regeneration. Finally there are also potential pharmacological avenues that may improve outcomes in nerve regeneration, such as geldanamycin and FK-506.

Keywords: Facial paralysis, paralysis, reanimation, gracilis

23.1 Introduction

The muscles of facial expression are perhaps the most obvious outward, involuntary expression of our internal emotional state. Impairment of its actions is not life-threatening, but will alter not only the appearance of the face at rest and in motion, but also, possibly, ocular and oral continence. Interestingly, these latter two are often not a significant issue in the pediatric population. The problem that most often brings the pediatric patient to the facial palsy clinic is in fact their inability to smile. Simply not being able to smile spontaneously at the appropriate time during a social interaction has a significant impact on the individual's psychological state and potentially also on the perception of others about the individual.

Pediatric facial palsy is relatively rare, and as with many pediatric conditions can be categorized into the congenital and acquired. Although facial palsy present at birth has an incidence of 2.1/1,000 in newborns, most are acquired and associated with birth trauma such as forceps use, and the incidence of actual congenital cases is closer to 0.24/1,000—around 90% of birth-injury-associated palsies show complete recovery. Facial palsy acquired after birth, while less common than in adults, is usually associated with an infective process and typically presents with an acute-onset facial paralysis, the most common

reasons being Bell's palsy, acute otitis media, and Lyme disease, and has an annual incidence of 6.6/100,000. The management of the patient should be within a multidisciplinary environment that includes ear, nose, and throat surgery, rehabilitation therapy services, psychosocial support, and a pediatric medical assessment.

23.2 Comparison with the Adult Population

- *The etiology of the disorder.* In congenital or developmental facial palsy, it is not possible to re-neurotize using the ipsilateral facial nerve. Furthermore, there may be no useful muscle units, and if dynamic reconstruction is required, the use of a functional muscle transfer is mandated. Also, in cases of bilateral facial nerve palsy even the contralateral facial nerve may not be available to use as donors. Importantly, facial palsy apparent at birth is not necessarily a “development” problem, but may be secondary to birth trauma, for example, following the use of forceps.
- *Psychosocial.* The ability of a child to be able to respond to a facial expression with their own (gestural coupling) is important for psychosocial development, and its loss may affect not just the perception of others about the child with facial paralysis, but also how the child understands others. On the other hand, patients who acquire facial palsy already have established social functioning. The psychosocial implications, therefore, of a facial palsy are potentially greater in the congenital than the generally older, acquired group.
- *Expectations.* The adult population with facial paralysis tend to have a lower expectation of what must be achieved (primarily focused on functional issues) and are often keen for less involved procedures to achieve more simple results, whereas the opposite is the case for the younger population—often driven by parental desire for a child who can integrate “normally” with their peers.
- *Peripheral and central neural plasticity.* Peripheral nerve healing in children is much more effective than in adults. Therefore, greater success can be expected in techniques for facial reanimation that depend upon it in children compared to adults. Furthermore, central plasticity is also better in the younger patient and is of particular merit in procedures in which a nerve other than the facial is used to innervate a reconstruction.
- *Co-morbidities.* In the elderly adult group with, for example, malignant parotid tumors, anesthetic limitations and the likely prognosis may limit which procedures are appropriate. In the pediatric population, the facial palsy may be syndromic and reanimation must be coordinated with other treatments and may be affected by them. For example, in hemifacial microsomia, the final aesthetic outcome will also be very dependent upon the craniofacial reconstruction, or where a future maxillary osteotomy may be needed, a cross-facial nerve graft that could be injured by such a procedure may be contraindicated.

23.3 Anatomy of the Facial Nerve and Muscles of Facial Expression

The first branches of the facial nerve once it leaves the stylomastoid foramen are the posterior auricular branches, which are motor to occipitalis, and branches to the stylohyoid and posterior belly of digastric. The nerve then has a short course between the foramen and the parotid gland through which it passes, separating it into deep and superficial lobes; despite this intimate relationship, the facial nerve provides no innervation to the parotid. As it passes through the gland, the nerve branches into the pes anserinus, and by the time it exits the parotid is classically said to do so in five major branches: zygomatic, temporal, buccal, marginal mandibular, and cervical (► Fig. 23.1). This is, however, an oversimplification and there are not only multiple branches, but also a significant degree of cross-innervation between branches.

The nerve is deep to the superficial aponeurotic system (SMAS), and it supplies the muscles of facial expression. The muscles are layered into four levels, and the branches of the nerve lie deep to all but the deepest layer (which consists of the buccinator, mentalis, and levator anguli oris). The muscles may be divided roughly into thirds: an upper group (in the supraorbital and periorbital region), a midface group, and a lower face group. The upper group (frontalis, procerus, and corrugator supercilii) originate laterally and superiorly and insert into the glabellar and supraorbital area. They function to animate the eyes and periorbital regions. With the exception of nasalis, the muscles of the midface and the lower face insert into the oral commissure and thus animate the lips (► Fig. 23.2). In 1973, Rubin described three basic types of smile (► Fig. 23.3). The first, the “Mona Lisa” smile, accounted for 67% of his subjects and was due to the dominant action of the zygomaticus muscles. The second, “canine smile”, accounted for 31% and was due to a greater dominance of the levator labii superioris. The third group was the rarest, the “full denture smile,” which

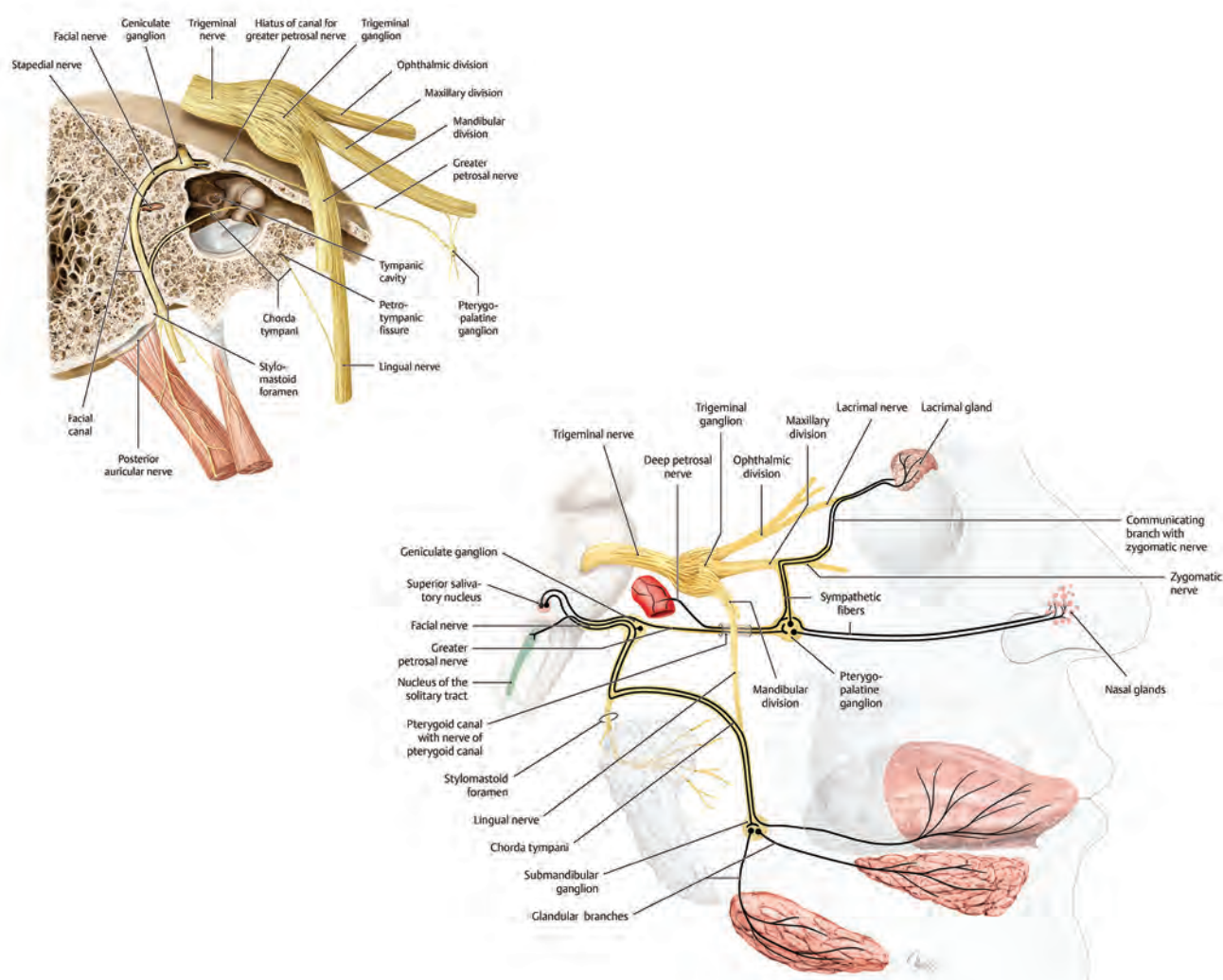


Fig. 23.1 The facial nerve nucleus is located in the brainstem, travels through the cerebellopontine angle, then courses intratemporally before exiting at the stylomastoid foramen. Each of the extratemporal facial nerve branches provides specific movement. (From THIEME Atlas of Anatomy, Head, Neck, and Neuroanatomy, © Thieme 2015, illustration by Karl Wesker.)

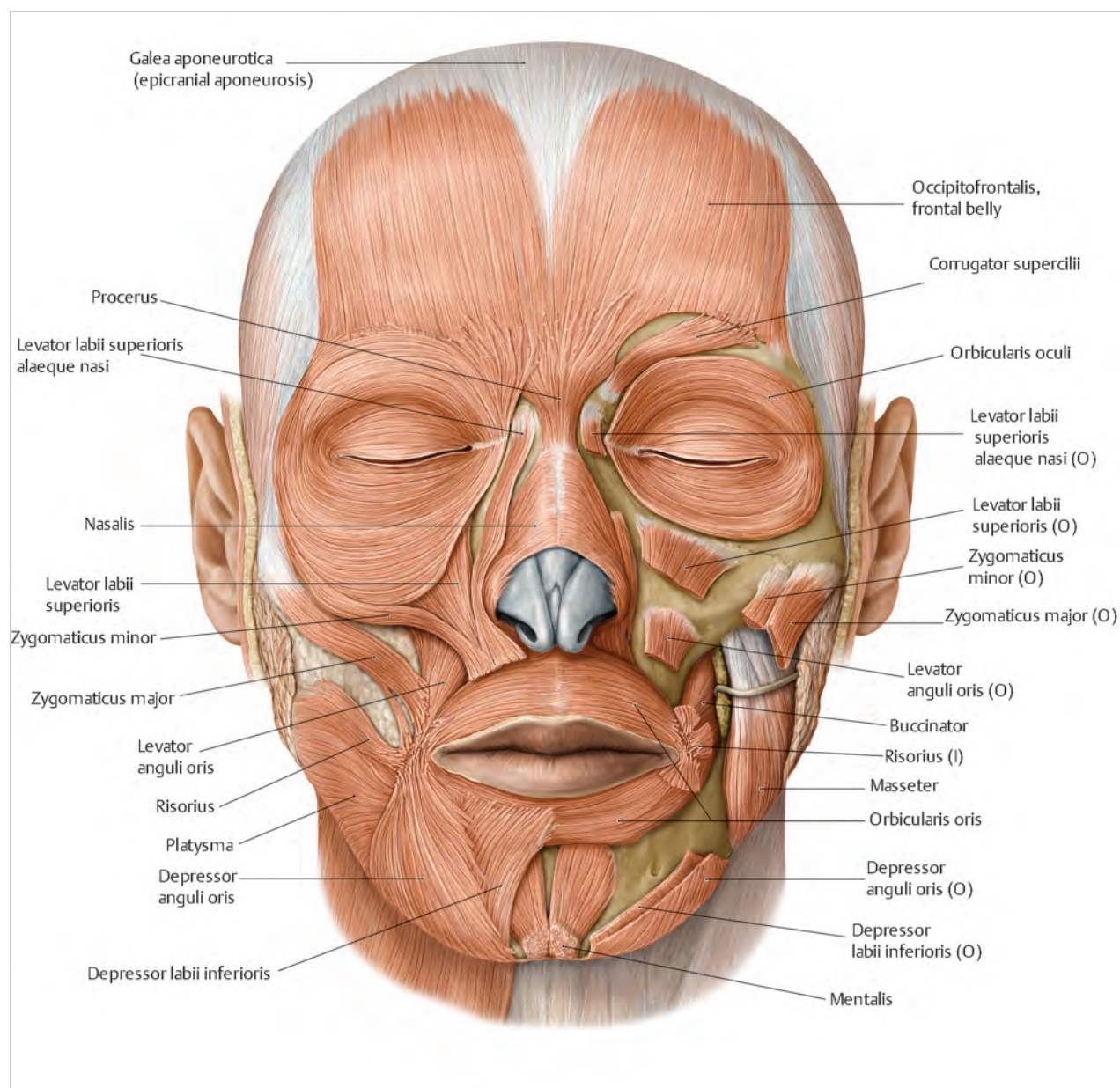


Fig. 23.2 The muscles of facial expression. (From THIEME Atlas of Anatomy, Head and Neuroanatomy, © Thieme 2010, illustration by Karl Wesker.)

accounted for just 2% of the study group and was felt to be due to roughly equal forces being applied to the lips from all the muscles inserting into them. We believe aiming for a line that mimics the zygomaticus muscles gives the best results.

At the nasolabial fold, the facial musculature integrates with the dermis. The consequence of this uniquely intimate relationship between the underlying muscle and the overlying integument is that relatively small movements have a significant effect on the overlying skin and soft tissues. Also, as a result, unlike in muscle transfers elsewhere, it is our experience that the insertion of the muscle should be to the dermis in order to effect the best results. Following denervation, the motor endplates at the neuromuscular junctions atrophy such that by 2 years following the loss of their nerve supply, they are not functionally re-neurotizable. Following facial nerve injury, therefore,

this defines the window of opportunity available to re-innervate the muscles.

23.4 Diagnosis and Assessment

It is important to recognize that facial paralysis is not necessarily a diagnosis in itself, but often a sign or symptom of an underlying condition. The assessment of the child with a facial paralysis is aimed at the following:

- Recognizing underlying or associated symptoms.
- Evaluation of the nature of the paralysis and its effects (and particularly any acute functional problems).
- Working out reconstructive goals.
- Working out reconstructive options.

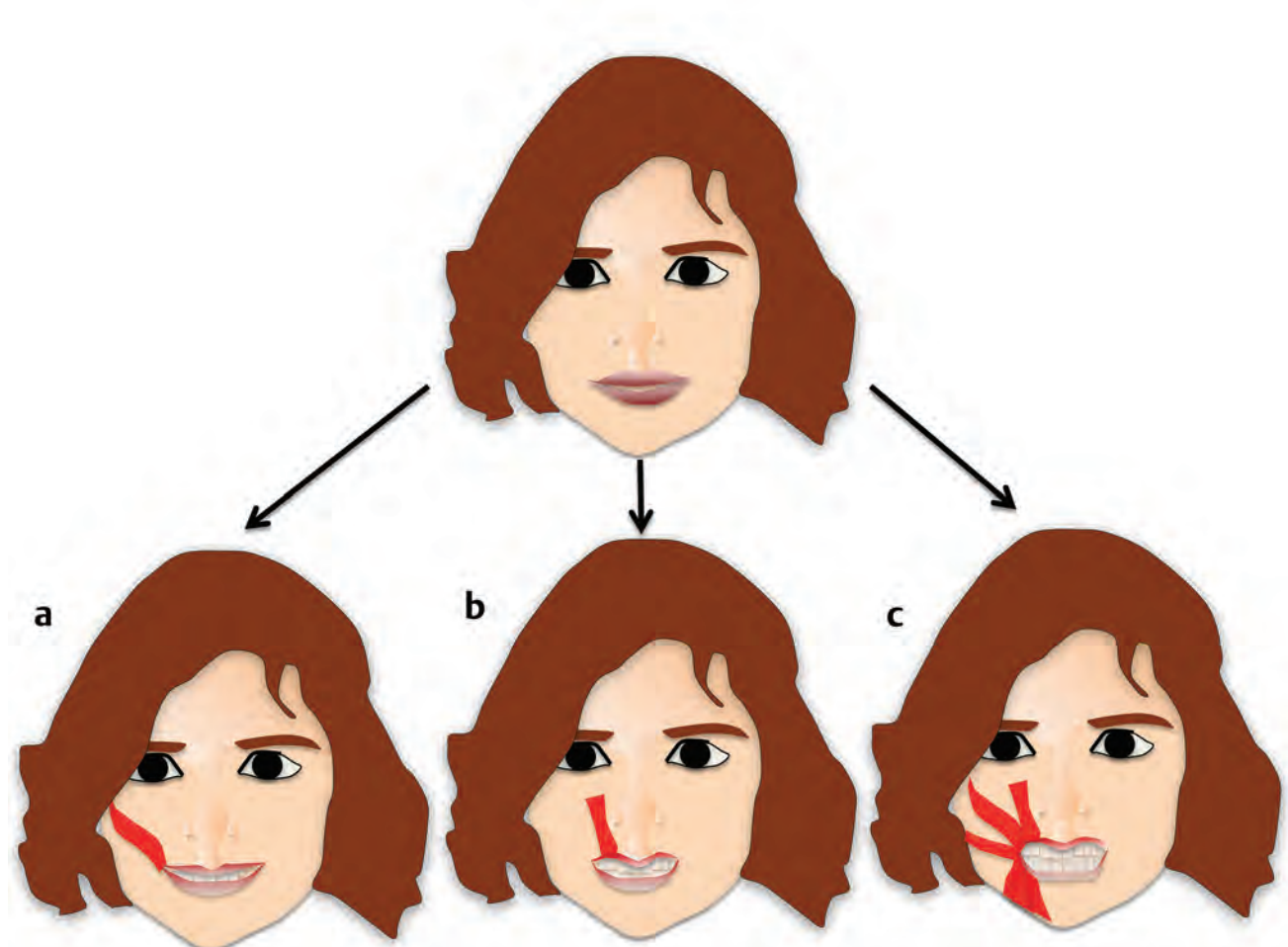


Fig. 23.3 The three smile types described by Rubin from his 1973 article. (a) The “Mona Lisa” smile, in which the primary movement is due to the action of the zygomaticus muscles. (b) The canine smile, where the dominant muscle acting is the levator labii superioris. (c) The full denture smile, which is due to comparably forceful contraction of all the muscles inserting into the lips.

Assessment is performed in a multidisciplinary environment including the surgeon, therapy services, a pediatrician, and potentially other specialties as required, such as ear, nose, and throat surgery. A targeted history and examination is performed to elucidate the likely cause of the facial palsy and therefore the prognosis and possible other medical problems, as well as therapeutic options. A list of the possible causes of the facial palsy is given in Box 23.1. In the history, the pertinent points are whether the palsy has been present since birth (and if so the type of delivery) or whether it was acquired, its duration, whether it is bilateral or unilateral, whether it is static, improving, worsening, or episodic, and other symptoms such as strabismus (as may occur in Moebius syndrome). If the child is old enough, information on the function of other parts of the facial nerve is also useful, such as whether there is reduced lacrimation or if hearing and taste have been affected. Furthermore, questions regarding functional problems such as epiphora and ocular trauma, speech impairment, and problems with feeding should be enquired about. Finally any history that may be suggestive of a specific neoplastic lesion should be aggressively pursued. The physical examination should focus on working out the level of the facial nerve injury, the branches affected, and the severity of the effect on the brow, periorbital

area, midface, and lower face (► Fig. 23.4, ► Fig. 23.5). Particular attention is paid to whether the eyes are affected, whether eye closure is complete, and if not the quality of the protective Bell's reflex, as the combination of an inadequate Bell's reflex and poor lid closure exposure is potentially sight threatening due to the resultant corneal exposure, though, ocular problems in the pediatric group are rare compared to adults. Other points of note on physical examination include any evidence of hemifacial microsomia, the position of the cupid's bow on attempted smile—in congenital cases this tends to be more centralized—the function of the muscles of mastication and tongue (given the motor nerve to masseter and the hypoglossal are potential donors), the vector of the unaffected lateral lip element on smiling, and a “gestalt” view of the face. In our experience, electrophysiology does not alter management. Imaging will rarely usefully visualize the facial nerve, but may be relevant to identify other potentially important pathologies such as space-occupying lesions. Through this process, one should be able to determine the following factors: *Is this congenital or acquired? What are the deficits? What does the patient/parents feel are the problems relating to appearance? What is the chronicity of this palsy? What are the available donors? What are the goals?*



Fig. 23.4 A young girl with a congenital right-sided facial palsy. In (a), she has been asked to smile. (b) The preoperative markings. N refers to the normal side and P to the paralyzed side. While there is clearly asymmetry at rest, it is most marked during the attempted smile. And while all branches of the facial nerve are affected, the asymmetry is most noticeable around the lips and in particular when the child is asked to smile. Also, of note, the eye is not as markedly affected as the mouth. The arrow on the right cheek in (b) corresponds to the vector of pull on the oral commissure that is felt, which would give the most similar result to that of the left side of the face seen in (a). The intended nasolabial crease is also marked in (b) with reference to the normal contralateral side.

Box 23.1 Summary of Conditions That May Cause Facial Paralysis

- Extracranial
 - Traumatic
 - Facial lacerations
 - Blunt forces
 - Penetrating wounds
 - Mandible fractures
 - Logogenic injuries
 - Newborn paralysis
 - Neoplastic
 - Parotid tumors
 - Tumors of the external canal and middle ear
 - Facial nerve neuromas
 - Metastatic lesions
 - Congenital absence of facial musculature
- Intratemporal
 - Traumatic
 - Fractures of petrous pyramid
 - Penetrating injuries
 - Larogenic injuries
 - Neoplastic
 - Glomus tumors
 - Cholesteatoma
 - Facial neuromas
 - Squamous cell carcinomas
 - Rhabdomyoma
 - Arachnoidal cysts
 - Metastatic
 - Infectious
 - Herpes zoster oticus
 - Acute otitis media
 - Malignant otitis externa
 - Idiopathic
 - Bell palsy
 - Melkersson–Rosenthal syndrome
- Congenital osteoporosis
- Intracranial
 - Iatrogenic injury
 - Neoplastic: benign, malignant, primary, metastatic
 - Congenital
 - Absence of motor units
 - Syndromic
 - Hemifacial macrosomia (unilateral)
 - Mobius syndrome (bilateral)



Fig. 23.5 A 5-year-old girl with a partial bilateral facial palsy (Moebius syndrome) with the right side being more affected than the left. In image (a), she is at rest and in (b) she is attempting to smile. As with the child in Fig. 23.4, the asymmetry is most marked at the oral commissure, with the rest of the face (and of particular note the eyes) being relatively unaffected.

23.5 Reconstructive Aims

Broadly speaking, reconstructive objectives fall into two main groups: amelioration of functional impairment and amelioration of the appearance. These two concerns can themselves be further classified hierarchically according to their potential consequences for the patient. This grading is to a degree arbitrary, but enables a structured approach to management planning.

23.5.1 Problems Relating to Physiological Functional Impairment

- **Ocular protection.** Though often not a problem in the pediatric group of patients, facial paralysis can lead to a significantly increased risk of corneal ulceration, which itself can lead to blindness, and in one study of Bell's palsy, over half of the affected patients developed signs of corneal ulceration at 1

year. This is a particular problem in the patient in whom there exists a poor Bell's reflex (the reflex upward movement of the globe on attempted lid closure). In addition to the risk of corneal ulceration, the combination of dry eyes due to corneal exposure and epiphora due to lack of apposition of the lower lacrimal punctum to the globe is quite distressing for patients.

- **Oral continence.** While in adults and older children oral sphincter incompetence may be socially embarrassing, it can actually lead to a failure to thrive in neonates—in particular if bilateral. Furthermore, speech development may be affected and dental decay accelerated. Though, again, these problems are rare.
- **Nasal continence.** Nasal airway obstruction can occur due to loss of functional musculature (nasalis and levator alaeque nasi). However, this is rarely an issue when unilateral and only potentially so when bilateral—in general, this is not a significant presenting complaint in the pediatric population.

23.5.2 Problems Relating to Appearance

This is what the majority of pediatric facial palsy patients present with—and specifically an inability to generate a smile that falls within the range of what the family perceive as “normal.” Addressing the appearance of facial paralysis may have not just an aesthetic, but a psychological and social impact too. Again, as with the functional issues, the goals pertaining to the management of the appearance of the paralyzed face may be stratified for convenience in the usual order of importance for the patients:

1. Alleviation of socially embarrassing sequelae of paralysis—drooling, an unblinking “staring” eye, or apparently sad face at rest.
2. Restoration of symmetry at rest.
3. Restoration of dynamic symmetry.
4. Restoration of spontaneous, dynamic symmetry.

Appreciation of the symmetry is vital here—as in unilateral cases, the affected side is abnormal *by comparison with* the unaffected side. Furthermore, this means that treating the asymmetry by treating the *unaffected* side as a balancing procedure, for example, with botulinum toxin or myotomy, may be a reasonable option.

Another consideration is synkinesis (the, unwanted, involuntary contraction of a muscle in association with the contraction of another). It is a problem in facial palsy due to aberrant reinnervation, and further negatively impacts on the appearance of the paralyzed face. Synkinesis is not as great a problem in the pediatric population as in the adult population, given the patients being managed in the pediatric facial palsy clinic are often those with permanent facial palsy following trauma or developmental dysgenesis (where there may not even be muscle which can be reinnervated aberrantly or otherwise), in whom spontaneous re-neurotization is unlikely. Therapeutic options for synkinesis include biofeedback therapy, botulinum toxin, and cross-face nerve grafting.

23.6 Nonoperative Management

Rehabilitation and therapy services are fundamental components of management. However, in children, the situation may be more problematic. This is for two reasons. First, children may be more challenging to engage with and a play-based approach is required. Second, there may be little chance of recovery (e.g., following tumor resection or a congenital anomaly) and so a more limited role for rehabilitation, and more of a role for the encouragement-adaptive techniques.

Many nonsurgical methods in pediatric facial palsy focus on the management of the exposed eye. Lubricating eye drops should be used regularly to prevent drying of the eye, and lower lid taping and night patching may also be used, although these are sometimes not well tolerated in children. Another extremely important issue is the recognition that at its heart, pediatric facial palsy is primarily a problem of how the child is seen to be interacting with the social group around them, and though operative treatments are largely focused on addressing this issue, there are other methods of helping with this that involve the input of nonsurgical staff, such as psychologists and social workers.

23.7 Operative Management

The ideal surgery for a facial nerve injury is a direct repair followed by nerve grafting of the defect. In theory, these may allow recovery of spontaneous, appropriately coordinated facial movements. However, particularly in pediatric facial palsy, this is often not possible because of the nature of the pathologies encountered. Reconstructive surgery is performed in recalcitrant facial palsy where spontaneous recovery is not expected. There are numerous procedures that have been described to manage facial palsy, and they have been previously classified according to whether they are “dynamic” (improve the active range of achievable movement) or static (which do not affect the active range of movement), and according to the area of the face that they are used to reconstruct. The dynamic procedures essentially either require neurotization of defunctioned muscle, the transfer of a functional muscle, or even the neurotization of a transferred muscle. On occasion, while a divided facial nerve or part of the facial nerve is awaiting appropriate neurotization to the distal stump (e.g., from a cross-face nerve graft), a “babysitter procedure” may be undertaken. In this procedure, another nerve (such as part of the hypoglossal either directly or via a nerve graft) can be transferred to that distal stump to prevent the deterioration of the affected musculature (i.e., loss of motor end plates) until such time as the cross-face nerve graft is ready for transfer (► Fig. 23.6). The “babysitter nerve” can then be detached from the facial nerve and the cross-facial attached. The various techniques are summarized in ► Table 23.1.

23.7.1 Brow Procedures

Lack of function of the frontalis renders the brow susceptible to ptosis, which may be aesthetically problematic or, if it obstructs the visual fields, functionally problematic too. In the pediatric population, unlike the adult population, this is not a significant problem, most likely due to the quality of the juvenile soft tissues. In general terms, the procedures that are considered for brow ptosis are the brow lift—endoscopic (least obvious scarring but least effective), bicoronal open, and open direct (most obvious scarring but most effective)—and contralateral balancing procedures, which may be temporary (e.g., with botulinum toxin) or permanent with a myotomy.

23.7.2 The Upper Eyelid

In facial palsy, the unopposed action of the levator can lead to lagophthalmos, resulting in corneal exposure, and potentially injury and scarring. Additionally, the efferent pathway of the blink reflex may not be functional. It is rarely a significant problem in the pediatric population. The most common procedure is countering the levator's action with a gold weight placed in the upper lid anterior to the tarsal plate (► Fig. 23.7). Classically a 0.8- to 1.2-g weight can be used. In older, more compliant children, an estimate can be made of which weight to use preoperatively by taping a trial weight to the upper lid preoperatively. Though there are many potential problems including its visibility, extrusion, and incorrect weighting, the relative efficacy, simplicity, and reversibility of the procedure mean that it is commonly used. Alterations to the technique include the use of

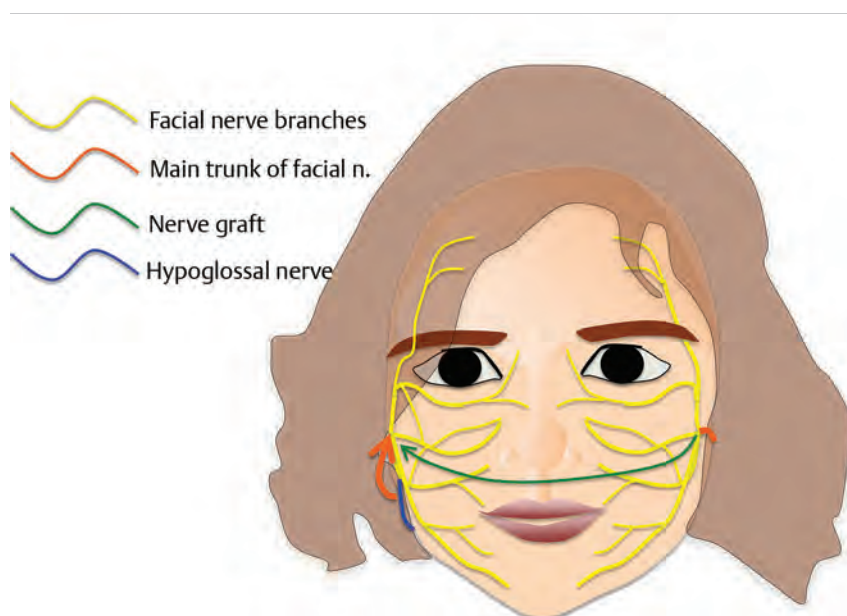


Fig. 23.6 Diagrammatic representation of a “babysitter” procedure in which the main trunk of the injured right facial nerve (DISTAL to the site of injury) is turned down and anastomosed end-to-side to the hypoglossal nerve temporarily neurotise the injured facial nerve and keep the motor end plates of the facial muscles intact until such time as axons from a cross facial nerve graft can reach the injured nerve. The arrows represent the direction of growth of the neurones.

Table 23.1 Summary of the various reconstructive procedures that may be used in facial reanimation

	Static procedures	Dynamic procedures
Brow	Direct excision brow lift	
	Open brow lift	
	Endoscopic brow lift	
	Contralateral weakening	
Upper eye lid	Weights	Palpebral springs
	Tarsorrhaphy	Temporalis transfer
	Levator botulinum toxin	Direct muscle neurotization
		Cross facial transfer
Lower lid		Free muscle transfer
	Static sling	Temporalis transfer
	Lateral canthoplasty	Direct muscle neurotization
	Medical canthoplasty	Cross-facial transfer
Nasal airway	Lid shortening	Free muscle transfer
	Cheek lateralization procedures	
	Tendon sling	
Commissure/upper lip	Septoplasty	
	Static slings	Direct muscle neurotization
		Direct muscle
		Cross facial transfer
		Regional muscle transfer
		Free muscle transfer

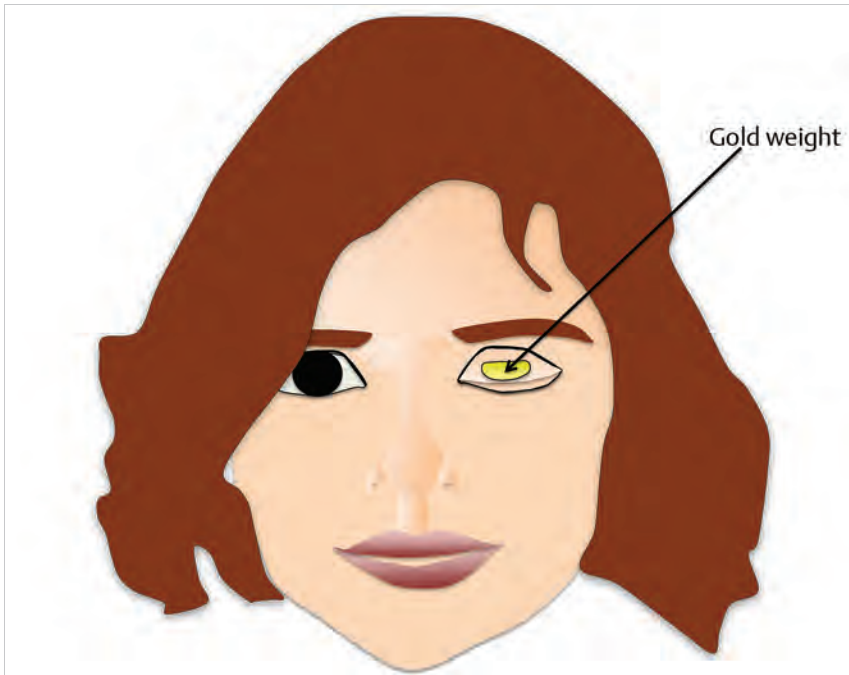


Fig. 23.7 A diagram showing the use of gold weights for the management of upper lid-related lagophthalmos. The weight is placed just anterior to the tarsal plate through an "upper-blepharoplasty" type incision.

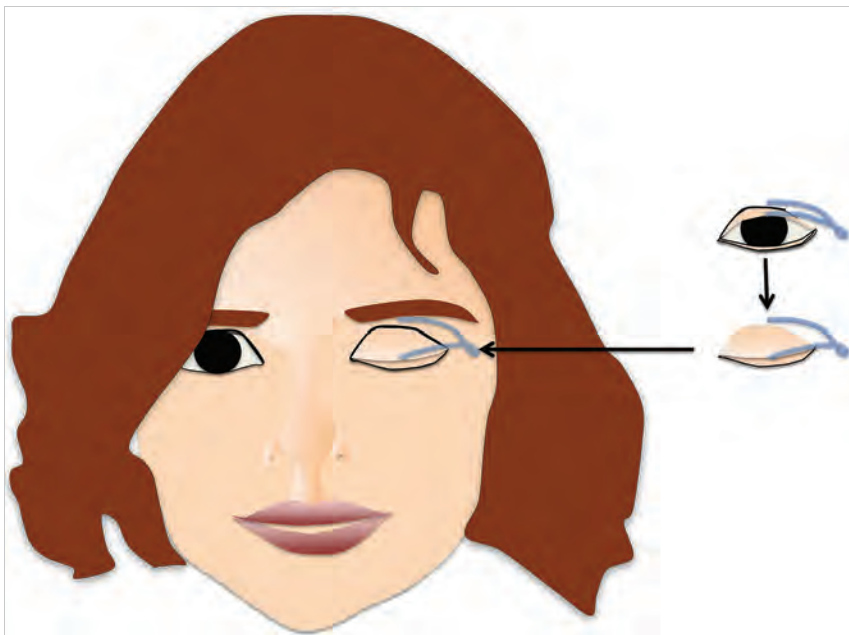


Fig. 23.8 A schematic demonstrating the spring technique by Morel-Fatio and Lalandie (1964). The spring (blue) is placed in the lateral canthal region. One arm of the spring pushes against the orbital roof and the other against the lower lid, enabling it to close.

smaller, lower profile platinum plates, and related techniques such as the use of magnetized and silicone-implantable devices, although these are less common.

Other static procedures that are more rarely used in the pediatric population include the use of a palpebral spring system (► Fig. 23.8) and tarsorrhaphy. The principle of tarsorrhaphy is to fuse the upper and lower eyelids by one of several different methods, reducing excursion and narrowing the palpebral aperture. A tarsorrhaphy is relatively simple and may be permanent, or where recovery or another definitive eyelid procedure is expected, temporary.

Of the dynamic procedures, a strip of temporalis muscle is dissected and transposed anteriorly, extended by means of

fascial or tendon grafts, and attached to the upper and lower eyelids, allowing voluntary (but not reflex) movement, controlled by temporalis contraction (► Fig. 23.9). More recently, dynamic "spontaneous and symmetrical" procedures have been developed, such as a free platysma transfer powered by a cross-facial nerve graft.

The use of botulinum toxin for the levator palpebrae superioris to counter the effect of the orbicularis may also be used as a temporary measure in order to allow corneal ulcers to heal.

Generally, contralateral balancing procedures are not appropriate for the upper eyelid as they would risk causing lid ptosis.

23.7.3 The Lower Eyelid

The lower eyelid is also not commonly a problem in children. While like the upper eyelid the problem is one impairing

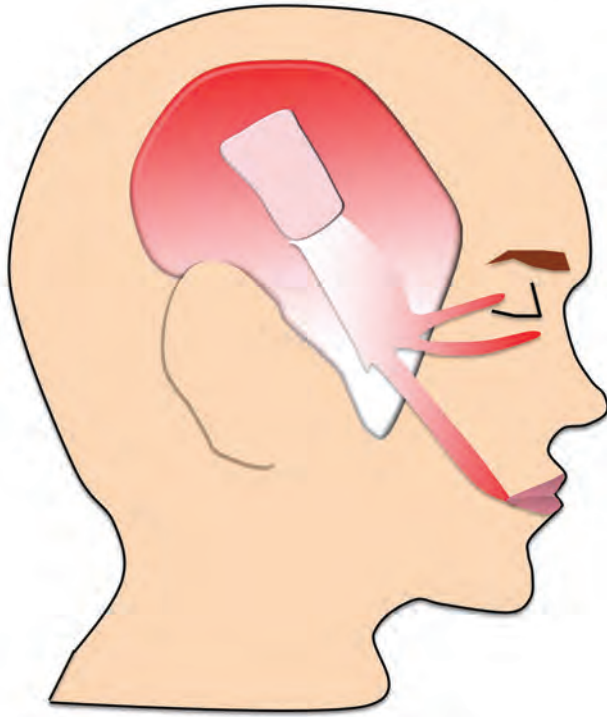


Fig. 23.9 Diagram adapted from Kazenstein describing the use of the temporalis transfer technique (1916). A segment of the temporalis muscle is turned down, split, and transferred to the eyelids and oral commissure.

closure of the eyelid, the etiology and effect of the problem are somewhat different. In the upper eyelid, the problem is one of unopposed levator action and inadequate motion, but in the lower eyelid, the “antagonist” is gravity and the problem is of inadequate support. It may be that the relative resilience of the pediatric population to lower lid problems is associated with their greater soft-tissue elasticity. The problem with lower lid paralysis is the potential for ectropion. The result is that due to the increased exposure of the eye, there is a feeling of dryness and also a paradoxical simultaneous epiphora, resulting from the loss of opposition of the lower lid lacrimal punctum to the globe of the eye.

In the event of lower lid ectropion, many of the dynamic procedures that can be used are similar to those for the upper lid—the temporalis transfer, cross-face neurotization procedures, and free tissue transfer. In our experience, though, static procedures—which essentially act to lend support to the drooping lower lid—are usually adequate. A tendon graft may be passed through the lower lid between the canthi and tightened to form a static sling helping appose the lower lid to the globe (► Fig. 23.10). Medial and lateral canthoplasties may also be performed. In the event of the primary problem being an excessively everted lacrimal punctum within the lower lid, a tarsoconjunctival ellipse may be excised from the medial lower lid to reposition the medial canthus. If the problem is primarily lateral, a lateral canthoplasty by a canthopexy or tarsal strip procedure may be performed, in which the lateral canthus is resuspended from the orbital rim.

23.7.4 The Nasal Airway

As a result of loss of ipsilateral soft-tissue support of the nose, the nasal airway may be prone to collapse. In unilateral cases, there is enough redundancy that the contralateral nasal airway is adequate to allow comfortable respiration, though if severe

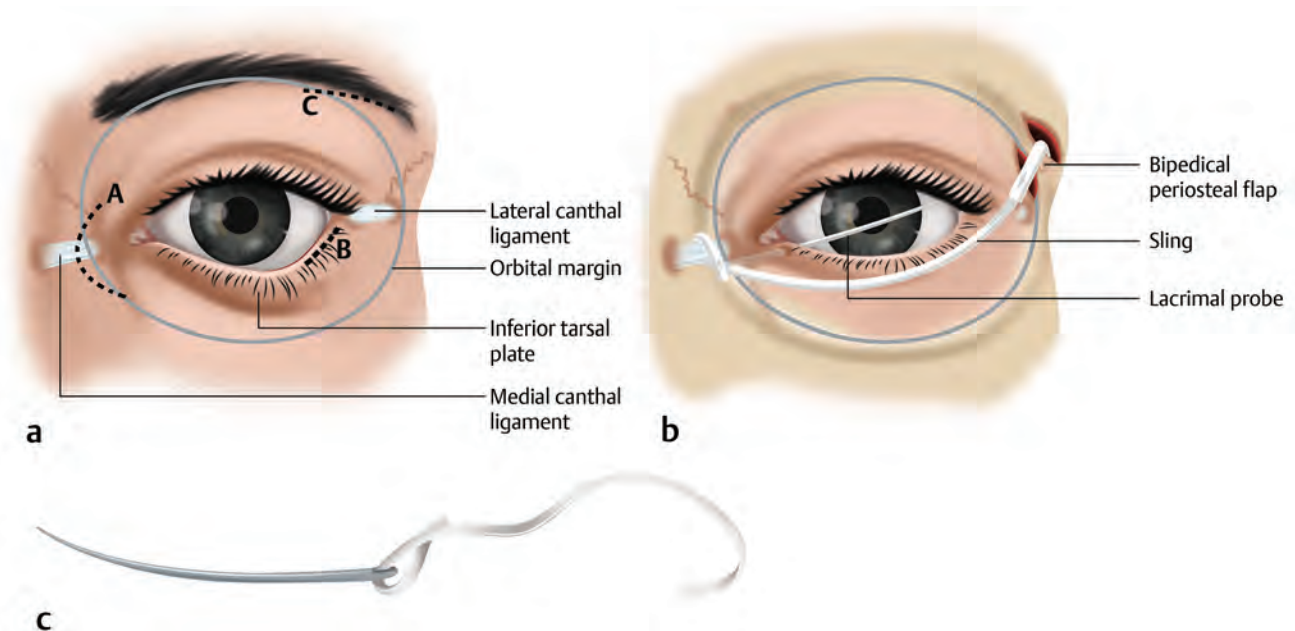


Fig. 23.10 (a) Incisions are located at A, B, and C. (b) The sling is positioned over the tarsal plate with a lacrimal probe in place to prevent damage to canaliculus. (c) A strip of tendon is passed through the eye of a Keith needle. (Adapted from Carraway and Manktelow 1999.)

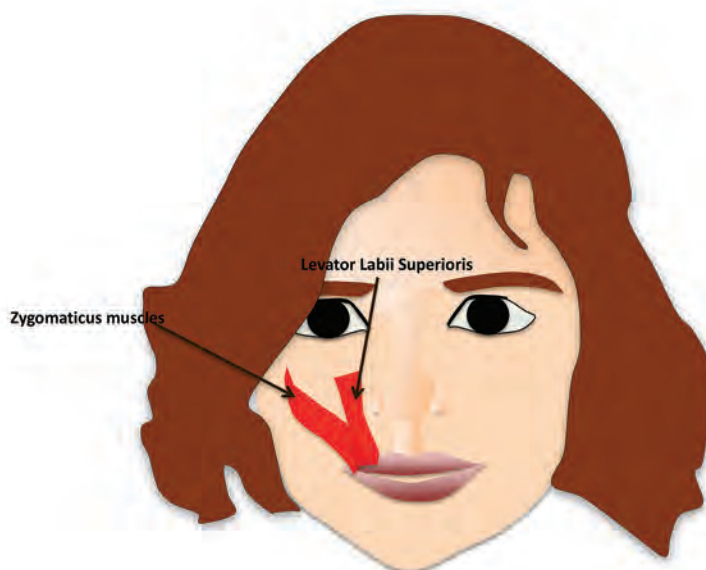


Fig. 23.11 Diagrammatic representation of the levator labii superioris and the zygomaticus muscles' action and giving their vector of pull on the oral commissure.

and bilateral it may be addressed with suspension procedures. Occasionally, if associated with septal deviation, it may benefit from septoplasty. However, in general, problems around nasal collapse are often addressed by secondary benefit of procedures that act to suspend the upper lip and oral commissure and so indirectly act on the nasal airway.

23.7.5 The Upper Lip and Oral Commissure

Within the pediatric population, the loss of midface symmetry and the “smile” is the most common reason for presentation, and the most common indication for surgical intervention. It is primarily as a result of the loss of function of the lateral and superior pull of the zygomaticus major and minor (“the Mona Lisa smile”), and levator labii superioris (“the canine smile;” ► Fig. 23.11). Over the years, many static and dynamic procedures have been developed to manage this problem. Static suspension of the upper lip and the commissure by means of tendon and fascia grafts and alloplastic materials such as Gore-Tex is a well-described option (► Fig. 23.12). For dynamic reconstruction, cross-facial nerve grafting may be done to residual distal nerve stumps where present, or as with the orbicularis oris for eyelid problems direct neurotization from the contralateral facial nerve is also an option.

Where no usable muscle is present, a muscle transfer is required. Local options include the use of the masseter, and perhaps one of the most well-known local options is the use of a temporalis transfer (► Fig. 23.13). In this technique, the temporalis muscle is mobilized from the posterior part of its origin in the temporal fossa and its insertion in the coronoid process is redirected under the zygomatic arch to the oral commissure.

Our preferred technique is the use of a two-stage procedure—a cross-facial nerve transfer followed by a free gracilis transfer.

In the first stage, a cross-face nerve graft is performed. Given that it is possible to use other local nerves to power the muscle transfer rather than a cross-face nerve graft, it is the authors' opinion that the great merit of a muscle powered by the facial nerve (albeit the contralateral facial nerve) is that it allows a truly emotionally spontaneous and bilaterally synchronous movement. The operation is performed as a two-team approach. The sural nerve is identified under tourniquet control at three locations by means of three short incisions just posterior to the lateral malleolus, in the midcalf and in the popliteal fossa. A specially designed nerve stripper is then used to delicately harvest the nerve, which is reversed prior to grafting. Simultaneously, the unaffected side of the face is exposed via a “face-lift” type incision, and the facial nerve is identified. The required branches are those that stimulate the desired “smile” on the unaffected side. These are then sharply divided (there is invariably adequate redundancy and crossover, so the nonparalyzed side's function is not impaired). The sural nerve graft is then either sutured or glued to these branches and tunneled to the contralateral side. The patient is then regularly assessed using Tinel's sign, and after a period of 4 to 6 months once the cross-face nerve graft has neurotized to the affected side, the second stage—the free gracilis transfer—may be performed.

In some cases, however, a cross-face nerve graft is not possible, for example, when there is a bilateral facial paralysis, and in this case the authors' second choice is a single-stage procedure in which the gracilis is innervated by the ipsilateral nerve to masseter. The gracilis is a type 2 Mathes and Nahai muscle flap (► Fig. 23.14), the dominant pedicle being from a branch of the profunda femoris. It is innervated by the anterior obturator nerve and its harvest causes minimal donor site morbidity. The original description by Harii of the flap was of the whole gracilis muscle, but the result was a bulky flap. This has subsequently been refined so that now it is only necessary to use a small limited segment of the muscle, using nerve stimulation to identify

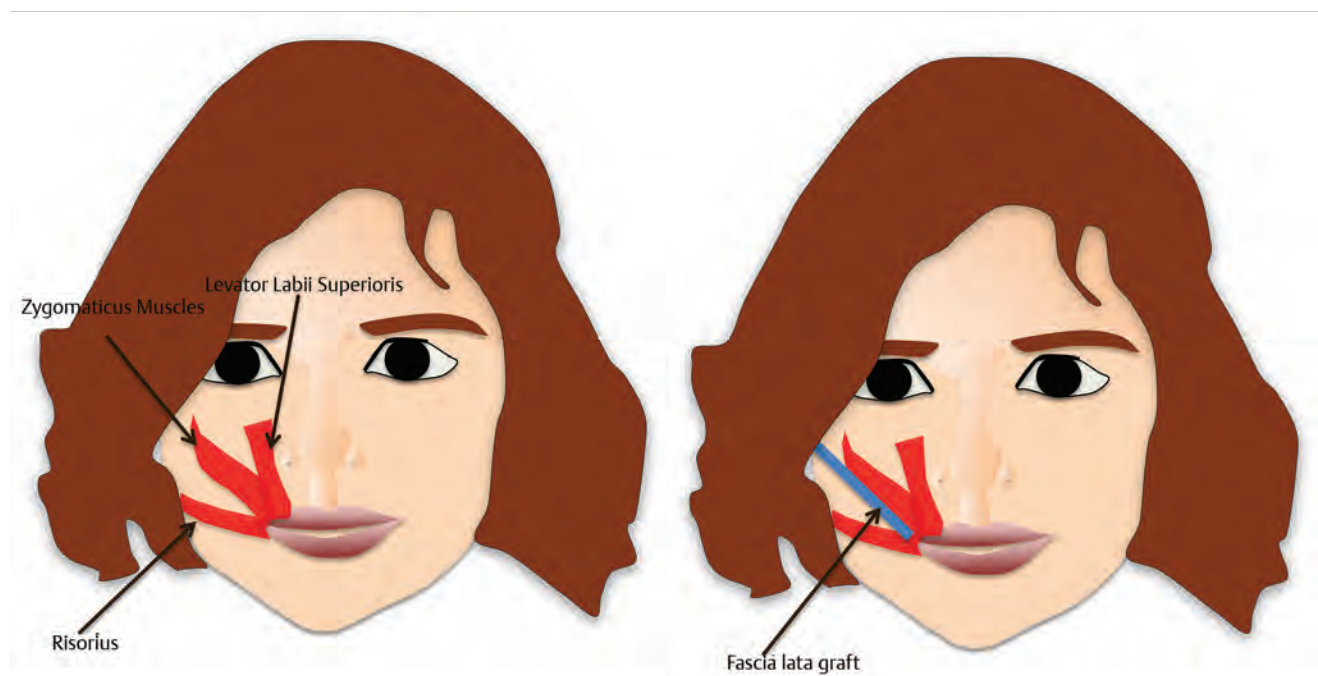


Fig. 23.12 Diagram based on schematics from Rose (2005) showing how a fascia lata graft (blue) running from the upper and lower lip to the zygomatic arch may be used to compensate for impairment of function of the levator labii superioris, the zygomaticus muscles, and the risorius.

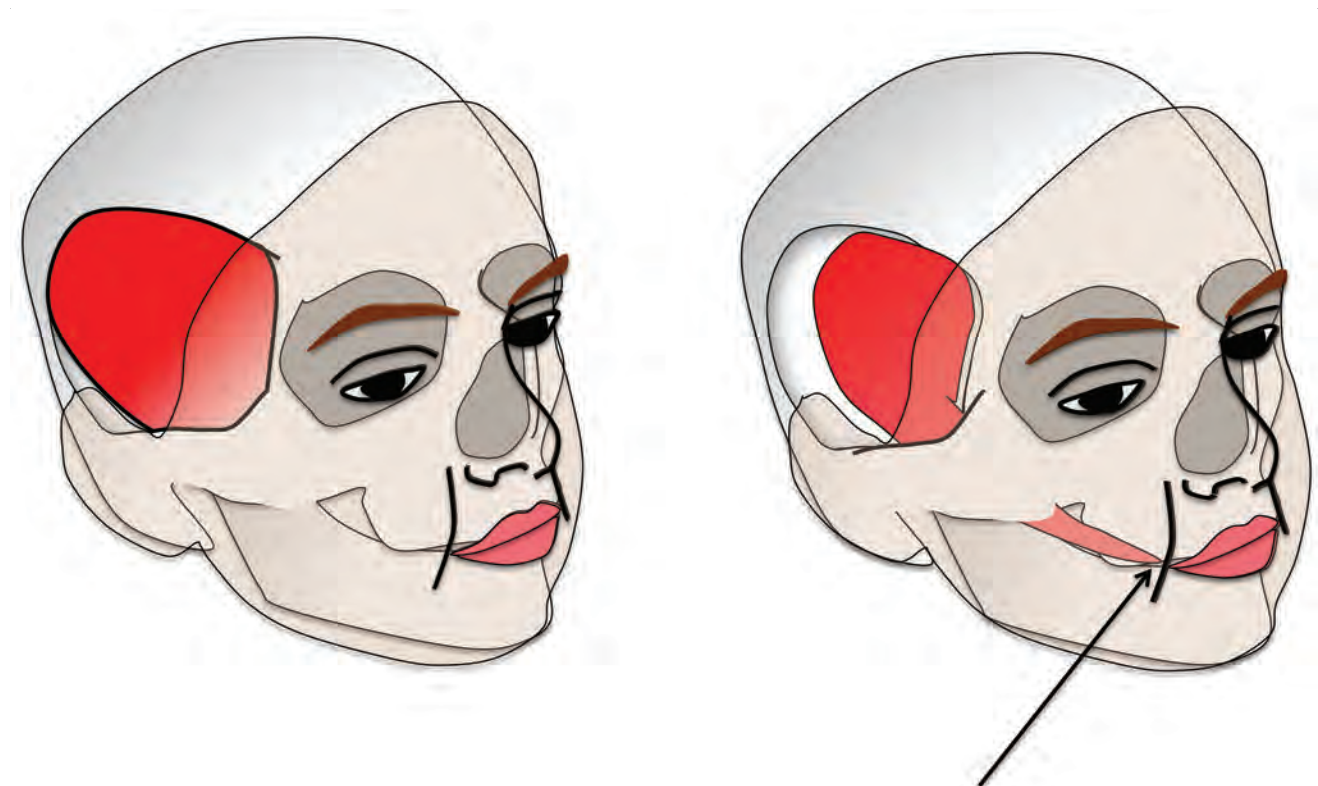


Fig. 23.13 Diagram of the temporalis lengthening myoplasty as described by Labbe (2009). The insertion into the coronoid is detached, and the posterior aspect of the muscle is detached from the temporal fossa, allowing the muscle to pivot and lengthening its reach so that the insertion can be passed beneath the zygomatic arch and can be attached to the oral commissure (arrow).

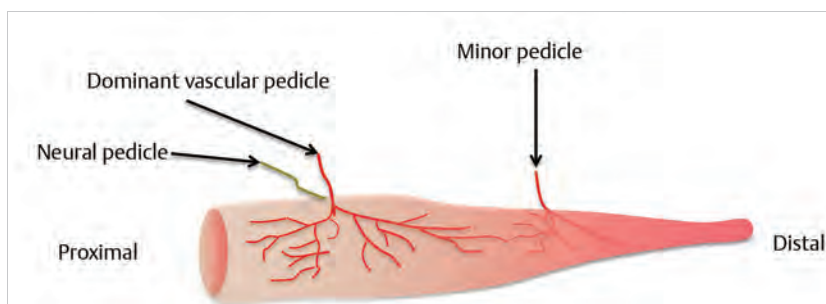


Fig. 23.14 Image based on radiographs of barium perfused specimens (in Cormack and Lamberty, *The Arterial Anatomy of Skin Flaps*) showing the vascular anatomy of the gracilis muscle. It is a Mathes and Nahai type 2 muscle, and the dominant pedicle and minor pedicles can be seen, as well as the neural supply from the anterior obturator nerve. Usually the dominant pedicle travels with two venae comitantes. (Adapted from Sharma et al 2016.)

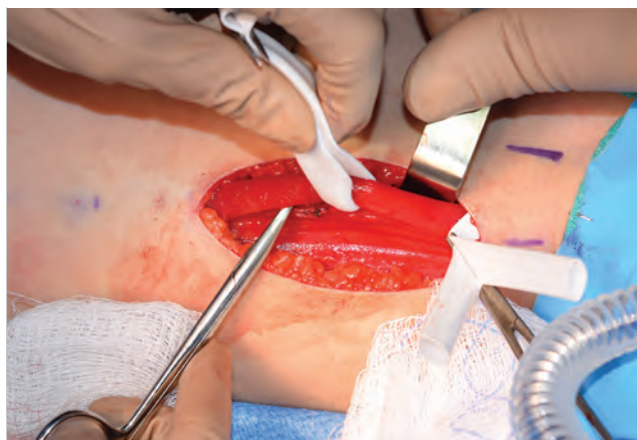


Fig. 23.15 Image of the gracilis in the right thigh. The segment of gracilis that stimulates most strongly with the nerve stimulator is identified and dissected from the main body of the gracilis, leaving behind up to 75% of the width of the gracilis.



Fig. 23.16 Photograph showing the externalized flap, giving an indication as to its size following the limited segment dissection.

the width of the muscle near the vascular pedicle most strongly stimulated by the nerve (► Fig. 23.15, ► Fig. 23.16). Key to the procedure is the preoperative marking of the patient to decide on the vector of pull that will be mimicked by the gracilis (► Fig. 23.17). This is challenging because normally there are multiple vectors from the multiple muscles attaching to the lip, but only one can be chosen for the gracilis. The muscle is harvested simultaneously with the preparation of the recipient site. The side of the face to receive the muscle is accessed via a “face-lift” approach in a subcutaneous plane. The buccal fat pad is removed to aid access to the preferred recipient facial vessels and to reduce bulk. Particularly careful attention is paid to the sutures placed to anchor the flap to the deep dermis around the oral commissure (as normal facial muscles may be expected to do) and with a line of pull that will tend to mimic a vector somewhere between the zygomaticus muscles and the levator labii superioris depending on the individual patient characteristics, and that is symmetric with the contralateral side, where appropriate. There should be a low threshold for adjusting them in the event of unsatisfactory placement (► Fig. 23.18). The muscle is then transferred and sutured in place anteriorly and inferiorly to the commissural anchoring sutures and posteriorly and superiorly to the temporal fascia (► Fig. 23.19) and revascularized, and the neurotomy performed to the chosen recipient. Other muscles also have been described in the literature, including the pectoralis minor and serratus anterior. A single-stage latissimus dorsi flap innervated from the contralateral side via the relatively long thoracodorsal nerve also has been described.



Fig. 23.17 Preoperative markings showing 1. the normal side (N) and the paralyzed side (P), the nasolabial fold on the right and a corresponding line on the left, the palpable facial vessels, dots around the right oral commissure where sutures will be placed, and an arrow demonstrating the intended line of pull.

23.7.6 Lower Lip

The muscles supplied by the marginal mandibular branch of the facial nerve are responsible primarily for lip eversion and depression, and these features are lost when the nerve is damaged. Static procedures include wedge excision (not indicated in the pediatric population) and the use of static suspension slings. In terms of dynamic reconstruction, as before, a cross-face nerve graft can be used to innervate the damaged nerve or

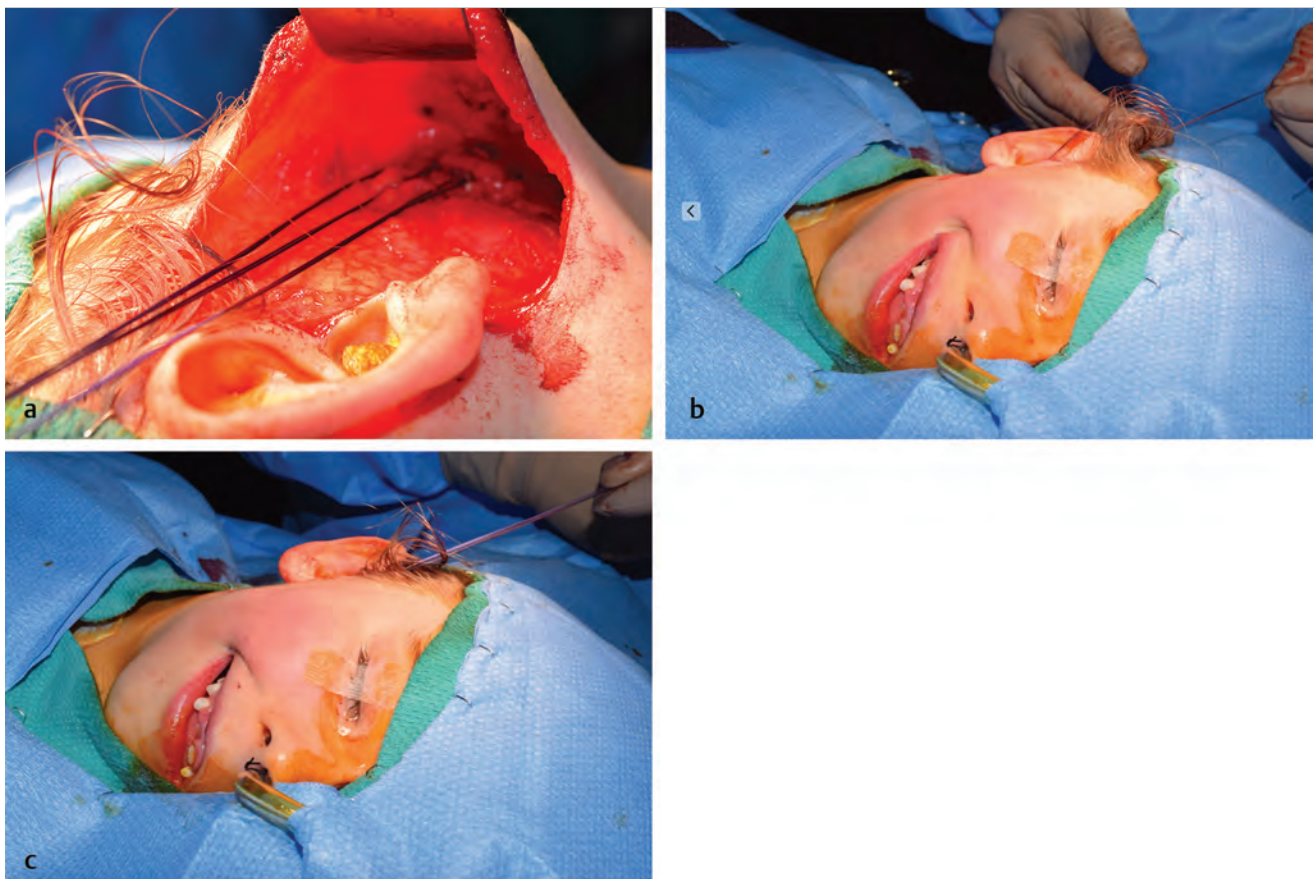


Fig. 23.18 Key sutures that will attach to the flap are placed at estimated ideal insertion site of the flap (a). The initial attempt results in an unsatisfactory result with an excessively lateralized nasolabial fold and lip eversion (b), and so the sutures are re-sited, resulting in a more acceptable outcome (c).

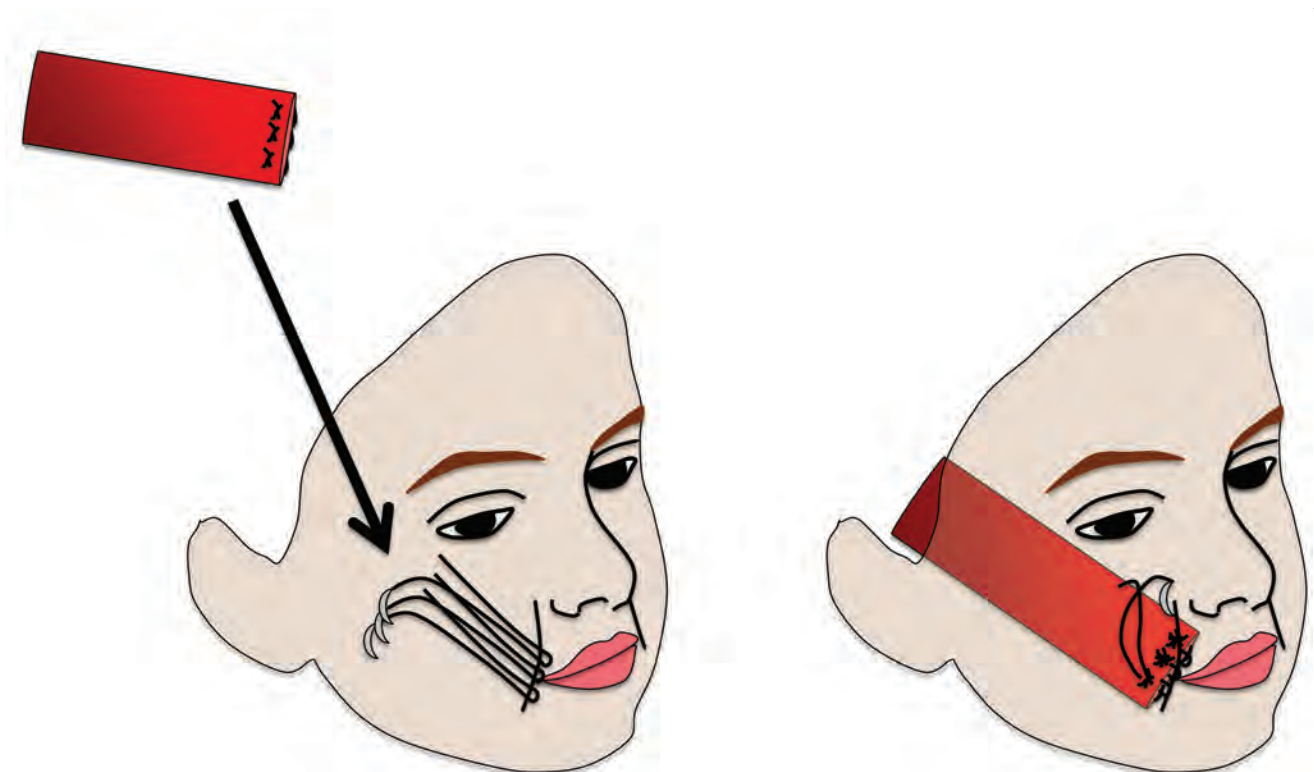


Fig. 23.19 The gracilis is transferred to the recipient site. The anchoring sutures are tied through the grasping "figure-of-8" sutures within the gracilis. The effect of these grasping sutures is to prevent the anchoring sutures pulling through the gracilis. The muscle is "parachuted" into place.

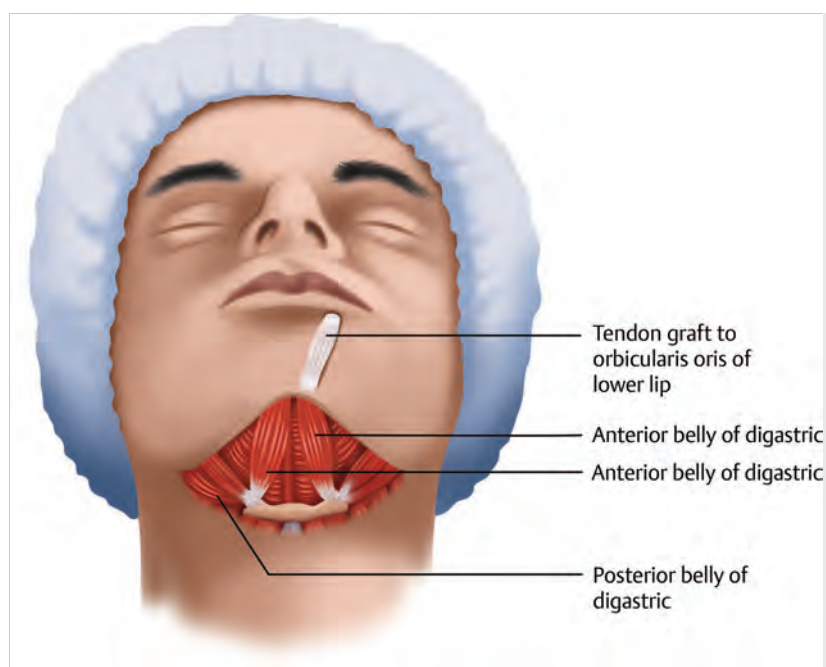


Fig. 23.20 An image of Edgerton's original picture (1967) describing the anterior belly of digastric transfer. The text reads: "When the lower lip is paralyzed, the asymmetry may be corrected by freeing the anterior belly of the digastric muscle from the mandible. A graft of fascia lata may be lashed to the muscle and split into two tails. Each tail is then threaded subcutaneously to loop about a bundle of the paralyzed lip muscle located near the commissure and just lateral to the midline. The ends of the fascia are returned to the neck incision, tightened with lip in slight overcorrection and sutured. As an alternative, the fascia may be sutured directly to the digastric tendon without freeing the muscle from the mandible. This latter method gives slightly less excursion of the lip."

even to neurotize the muscle directly. Regional muscle transfers have also been described—in particular the anterior belly of digastric (► Fig. 23.20), which is supplied by the trigeminal nerve, though neurotizing the transferred anterior belly of digastric with a cross-facial nerve transfer from the contralateral marginal mandibular nerve to give spontaneous facial nerve-dependent function has been described. Finally, contralateral balancing procedures may be performed—a myotomy and botulinum toxin injection.

23.8 Complications

In general, outcomes following free gracilis transfer are very good, with the vast majority of patients achieving an improved facial symmetry and a spontaneous smile (where a cross-face nerve graft has been used); the excursion achieved with the contralateral face can be 75% of that of the ipsilateral side. The complication rates are low, and in our unit over a 30-year period, only 1.6% of cases from 256 free gracilis transfers did not show an improvement in lateral lip excursion (i.e., either neurotization of the free tissue transfer failed) and would require a complete revision of the reconstruction, and only 3.9% required a minor revision. Very importantly, there is also now good evidence that facial reanimation surgery makes not only an objective, metric improvement in facial appearance, but also a subjective one in terms of the perception of others regarding the patient.

The management of suboptimal results includes the necessity for trimming of the muscle or subcutaneous recipient tissue in an excessively bulky flap. Incorrect positioning or slippage of the muscle along anchoring sutures is much more difficult to address. On occasion, these may be adjusted by repositioning, plication, or by adjustment with slings. However, these fine adjustments are often more difficult than they may appear and

sometimes the only solution is a complete revision of the flap. This has significant implications for the patient and their family and must be discussed with them at length. Other procedures that may become necessary include contralateral balancing operations.

23.9 Conclusion

The management of pediatric facial palsy begins with the accurate multidisciplinary assessment of the deficit and acute problems, the cause and associated problems, the likelihood of spontaneous recovery, and the psychological and social impact on the child and their family. After the management of acute issues (which are uncommon), once there is a decision to operate, an assessment is made of the surgical options. Which one is used depends on the exact deficit, but in general in the pediatric group the problem is the inability to smile. Our preferred option is a two-stage procedure in which the first stage is a cross-face nerve graft and the second stage a free gracilis transfer. This is gone through in detail with the family, and the decision to proceed is made together with them. The outcomes are generally good, with the vast majority of patients who have received the two-stage procedure achieving improved facial symmetry, developing an ability to smile spontaneously. It also achieves very satisfactory results for those receiving free gracilis transfers innervated by the motor nerve to masseter (► Fig. 23.21), and the complication and revision rates are relatively low.

In terms of the future, recent evidence from the rat model suggests that "supercharging" the nerve graft with a sensory nerve at the time of grafting can improve nerve regeneration. Brief periods of electrical stimulation of a proximal nerve stump can also enhance nerve regeneration. Finally, there are also potential pharmacological avenues that may improve outcomes in nerve regeneration, such as geldanamycin and FK-506.



Fig. 23.21 Before and after bilateral gracilis muscle transfers innervated by the motor nerve to masseter.

23.10 Key Points

- Management of pediatric facial palsy should be performed in a multidisciplinary environment including and collaboratively with the patient family.
- Assessment is to evaluate the possibility of sinister causes, the chances of spontaneous resolution, the deficit in terms of function and appearance and reconstructive options.
- The primary problem in children is the loss of an emotionally spontaneous smile caused by paralysis of the lateral commissure of the lip.
- There are many surgical options. At The Hospital for Sick Children in Toronto our preferred option for the lateral lip is a two-stage procedure: cross-face nerve grafting followed by gracilis transfer.

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24 Scalp, Cheek, and Neck Deformities

Arin K. Greene

Summary

Pediatric scalp, cheek, and neck deformities include: cutis aplasia, alopecia, hemifacial atrophy, and webbed neck. Skin lesions and trauma also may require reconstruction of these areas. Intervention is based on the type of deformity and the age of the patient.

Keywords: cutis aplasia, hemifacial atrophy, webbed neck, Turner syndrome, pediatric

24.1 Introduction

Reconstruction of pediatric scalp, cheek, and neck deformities can be performed by several methods. Scalp defects may be congenital (e.g., cutis aplasia), but more commonly result from extirpation of lesions or trauma. The most frequent congenital malformation of the cheek is hemifacial microsomia (covered in another chapter), followed by progressive hemifacial atrophy. Neck lesions such as branchial cleft anomalies and thyroglossal duct cysts usually are managed by pediatric general surgeons or otolaryngologists. Plastic surgeons are asked to improve soft-tissue neck deformities, such as webbing associated with Turner syndrome.

24.2 Diagnosis

Diagnosis of scalp, cheek, and neck deformities is made by history and physical examination. Imaging rarely is necessary, but may be indicated for cutis aplasia or hemifacial atrophy to determine if an underlying osseous abnormality exists. Occasionally, histopathology aids the diagnosis of scalp lesions.

24.3 Nonoperative Treatment

The primary morbidity of scalp, cheek, and neck deformities is psychosocial. Although cutis aplasia may require immediate operative intervention, other disorders can be observed. Timing of intervention typically involves three periods: (1) infancy, (2) between 3 and 4 years of age, and (3) late childhood or adolescence. Large lesions of the scalp are best removed during infancy because scalp redundancy at this time facilitates extirpation and reconstruction. If a deformity is likely to cause decreased self-esteem, then improving the condition between 3 and 4 years of age should be considered. Because long-term memory and self-esteem begin to form at approximately 4 years of age, correcting a deformity at age 3 often is desired. Some parents prefer to wait until children are old enough to decide whether or not they would like to have an operation, especially if the deformity is minor. Patients typically do not request a procedure until late childhood or early adolescence because before this time the fear of an operation outweighs their desire to improve a deformity.

24.4 Operative Treatment

24.4.1 Scalp

Cutis Aplasia

Cutis aplasia is a rare condition (1/5,000 newborns), and includes absence of scalp soft tissues, bone, and/or dura. (► Fig. 24.1). The disorder can affect any area of the body, but the scalp is the most common site (84%). The defect usually is along the sagittal suture and the area is covered with a thin membrane. One-fourth of patients have an underlying osseous or dural defect and thus are at risk for infection, venous thrombosis, and/or hemorrhage. Because cutis aplasia may be associated with syndromes and other anomalies, patients should be evaluated for additional medical conditions.

Management of cutis aplasia is based on the depth of the defect. If the bone is intact, superficial tissues are treated with local wound care and the area heals secondarily. Alopecia can be improved at a later date. If a cranial defect is present with exposed dura, the condition is life-threatening. Operative intervention is necessary within the first few hours after birth to prevent desiccation of the dura, meningitis, thrombosis of the sagittal sinus, and/or hemorrhage from exposed veins. If possible, the area should be closed by wide subgaleal undermining of scalp flaps. However, most defects are too large to close linearly and require a split-thickness skin graft over the dura. The thin graft is harvested from the gluteal area and is meshed. It is important to graft the dura before an eschar has formed. Once an eschar has developed, it is difficult to remove and the child is at risk for life-threatening hemorrhage and dural injury from debridement. If an eschar is present, it is best elevated off the dura using hydrodissection. Rarely, cutis aplasia can result in exposed brain without dura; the neonate requires emergent flap coverage of the area.



Fig. 24.1 Infant with cutis aplasia that healed secondarily. Decreased hair density can be improved with serial excision or tissue expansion.

Because osseous defects spontaneously heal in infancy, cranioplasty is not performed. Most bone gaps will close and rarely require secondary reconstruction. If a large osseous defect remains, it can be filled with particulate bone graft any time after 18 months of age when the dura no longer has the ability to create new bone.

Following closure of the soft-tissue defect, alopecia can be managed electively. Because the infant scalp has significant redundancy during infancy as the brain is rapidly enlarging, serial excision of the scar or graft should be considered starting at 6 months of age. If the area is not amenable to serial excision, then tissue expansion can be performed later in childhood. Small areas of alopecia can be observed until the child is older to determine if it will cause psychosocial morbidity.

24.4.2 Skin Lesions

Because most brain growth occurs during the first year of life, the scalp has unique redundancy during this period. Consequently, large skin lesions are best removed during infancy to facilitate the procedure and to achieve the most favorable outcome. I begin excision at 6 months of age when the risk of anesthesia is equivalent to an adult. If the lesion is very large and will take multiple serial resections, I perform the first resection at 3 months of age (► Fig. 24.2). The operations are performed at 6-week intervals when the wound has achieved 80% of its strength, and the adjacent skin has had sufficient time to relax. In some cases, the incision line cannot be completely closed

after widely undermining skin flaps and performing galeal scoring. Small openings (e.g., 1–2 cm) can be left open to heal secondarily.

Most scalp lesions can be removed by serial excision. However, some wounds require skin graft reconstruction if (1) the lesion is large, (2) the child has minimal scalp laxity, (3) the defect is in an area where it is difficult to advance skin flaps from two directions (e.g., ear), and (4) the lesion is unable to be partially excised because suturing remaining diseased tissue together under tension is problematic (e.g., neurofibroma, vascular malformation; ► Fig. 24.3). A split-thickness skin graft contracts, giving a wound that is approximately one-third smaller than the original defect. The skin graft then can be removed later by serial excision or tissue expansion.

Another option to reconstruct large defects following lesion removal is to expand the scalp prior to excision. This is a reasonable approach for nevi, but should not be performed for vascular lesions (e.g., neurofibroma, vascular anomalies) because tissue expansion can stimulate the disorder by increasing neovascularization; I prefer to remove the lesion, obtain a smaller abnormality with a skin graft, and then reconstruct the area secondarily with tissue expansion. I attempt to avoid tissue expansion whenever possible because it is traumatic to patients/families and is associated with significant complications (e.g., infection, extrusion). However, tissue expansion is the only alternative to reconstruct hair-bearing scalp for large areas not amenable to serial excision.



Fig. 24.2 Giant congenital nevus of the scalp. To take advantage of scalp laxity in infancy, serial excision of the nevus was initiated at 3 months of age. (a) Initial appearance with outline of planned resection. (b) Following first-stage excision. (c) Nevus is completely removed after four stages.



Fig. 24.3 Large neurofibroma of the scalp. Serial excision was not possible because the tumor could not be sutured to itself under tension. The wound was too large to be allowed to heal secondarily. Consequently, the lesion was removed in one stage and reconstructed with a skin graft. A thin split-thickness graft was used to allow maximal contraction of the wound to facilitate later serial excision of the graft or tissue expansion. Placement of tissue expanders initially was not chosen because of the higher risk of complications, potential stimulation of the tumor, and coverage of a larger wound compared to reconstruction of a contracted skin graft. (a) Preoperative appearance. (b) Following extirpation of the lesion. (c) Split-thickness donor site. (d) Bolster dressing placed. (e) Complete skin graft take. Note that part of the wound was closed linearly. (f) Six weeks postoperative appearance; the area of the skin graft is significantly smaller than the initial wound.

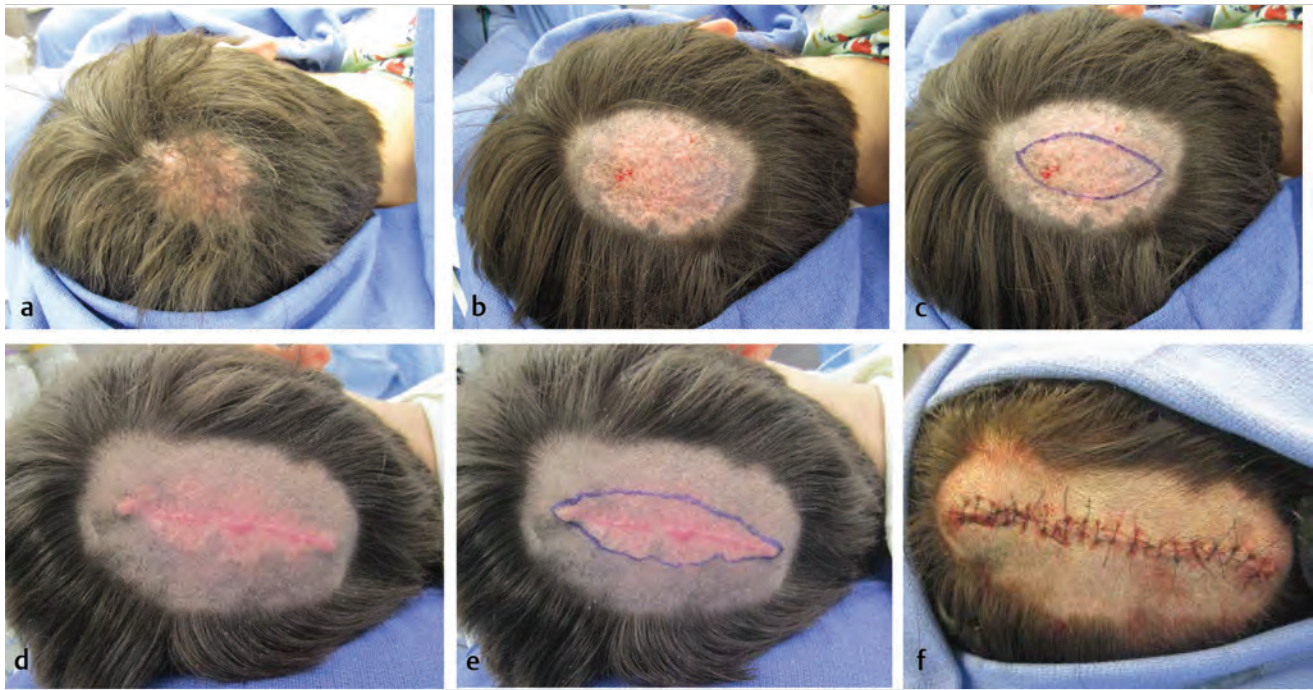


Fig. 24.4 Male child developed alopecia following a scalp infection. The condition was improved by serially excising the area of decreased hair density. (a,b) Preoperative appearance. (c) Outline of first stage excision. (d) Prior to second-stage excision. (e) Markings for second-stage resection. (f) Following final excision.

24.4.3 Alopecia

Alopecia can be improved by three methods: (1) serial excision, (2) tissue expansion, and (3) hair transplantation. If adequate scalp is present on either side of the alopecia, then the area can be improved by serial excision of the tissue with poor hair density (► Fig. 24.4, ► Fig. 24.5). Wide subgaleal undermining and scoring are necessary. If the area of alopecia is not amenable to serial excision, then tissue expansion is required (► Fig. 24.6, ► Fig. 24.7). I place the ports internally and patients return each week for saline expansion. Hair transplantation is efficacious for small defects involving the anterior hairline or for scars that are visible and cannot be narrowed any further (► Fig. 24.8). Donor scalp is harvested from the occipital region, micrografts are made under $3.5\times$ loupe magnification, and the follicles are transplanted into the recipient site.

24.4.4 Trauma

Traumatic wounds of the scalp are rarely problematic. Small areas can be allowed to heal secondarily. Larger lacerations should be closed, if possible, to prevent a wide scar that may cause visible alopecia (especially in males who wear their hair shorter). If a defect cannot be closed linearly, it is best to allow the area to contract and heal secondarily; the scar can be improved later. Acute rotational flaps, skin grafts, etc., should be avoided. If a large area of exposed cranium is present, it can be grafted over the periosteum to obtain a stable wound that can be reconstructed later. If the periosteum is not present, the ectocortex may be burred to bleeding endocortex, treated with negative pressure wound therapy, and the subsequent granulation tissue will accept a skin graft (► Fig. 24.9).



Fig. 24.5 Adolescent female with alopecia from a burn injury. (a,b) Preoperative appearance. (c) Intraoperative view following staged excision of scar. (d) Postoperative appearance following the second-stage excision of alopecia.

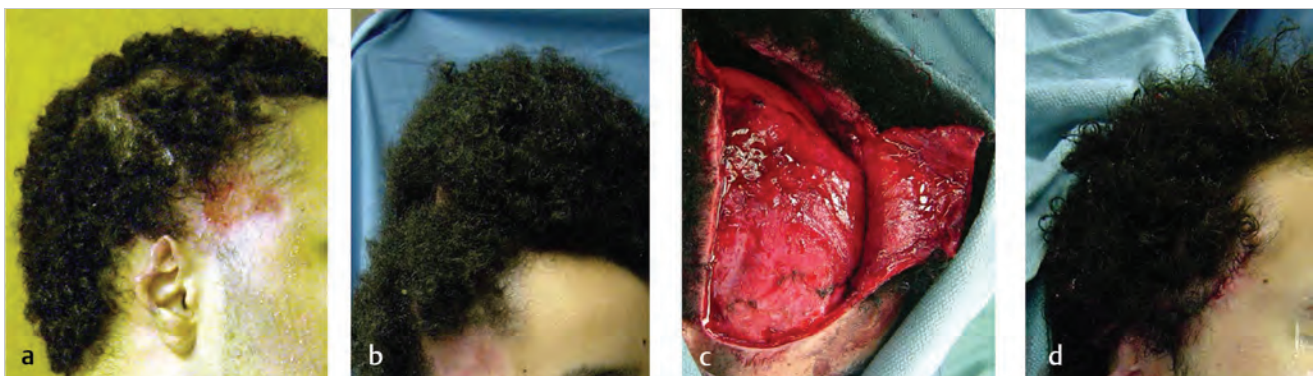


Fig. 24.6 Adolescent male who suffered a traumatic injury to his ear and scalp. Because the area of alopecia was large and located along the hairline, it was not a favorable location for serial excision or hair transplantation. (a) Preoperative appearance. (b) Tissue expanded scalp. (c) Scalp flap following removal of the tissue expander. (d) Inset of expanded scalp flap.

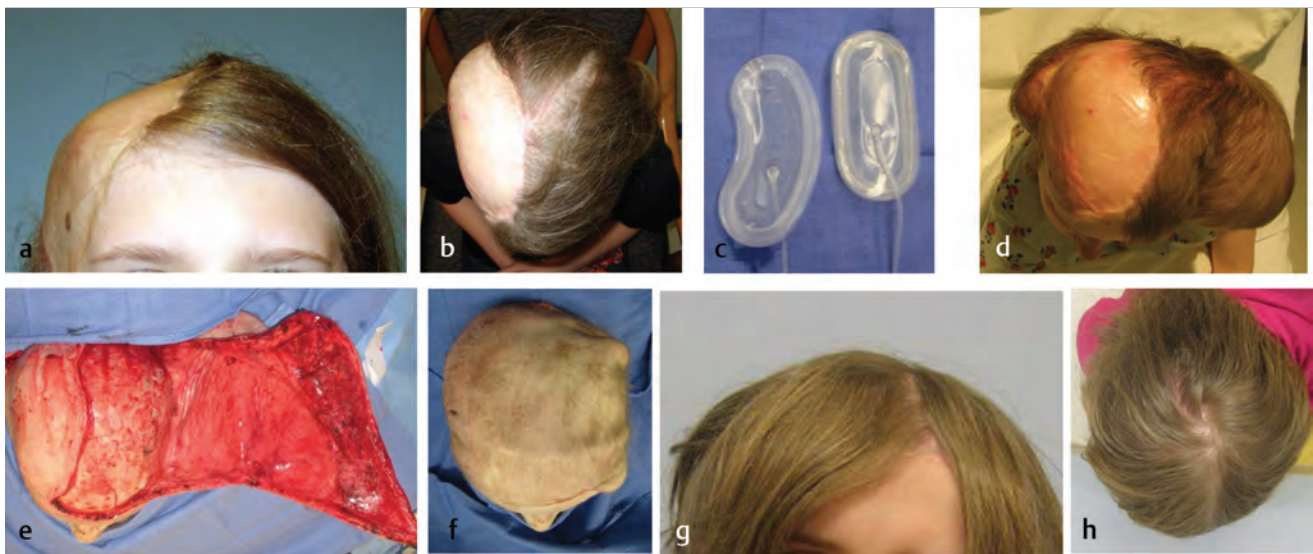


Fig. 24.7 Female child who underwent resection of a skin lesion reconstructed with a skin graft by another surgeon. (a,b) Preoperative appearance. (c) Two tissue expanders were placed. (d) Following expansion. (e) Expanded scalp flap. (f) Following inset of flap. (g,h) Postoperative result.

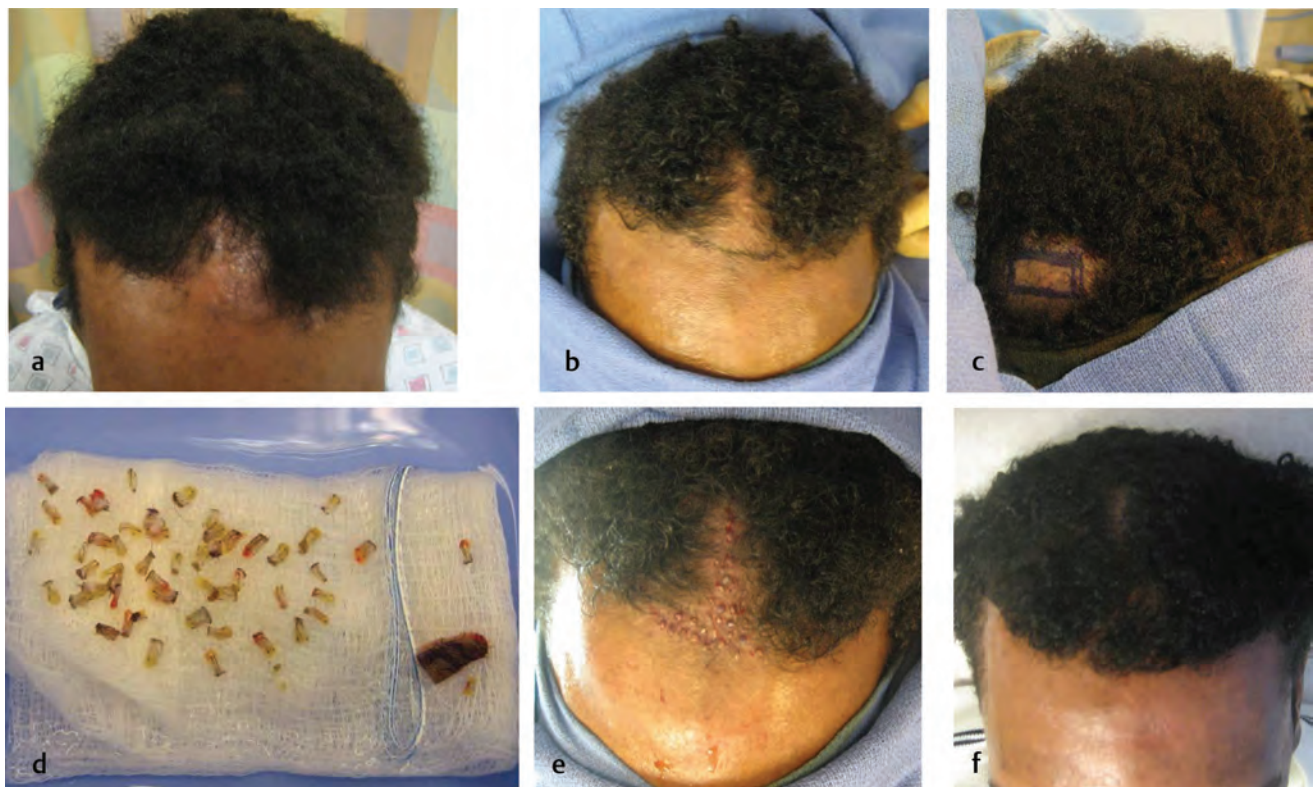


Fig. 24.8 Adolescent male with scar alopecia following trauma. Because the area was small and located along the hairline, it was managed with hair transplantation. (a,b) Preoperative appearance. (c) Donor hair harvested from the occipital scalp. (d) Micrografts prior to transplantation. (e) Intraoperative view of grafts in the recipient site. (f) Postoperative appearance.

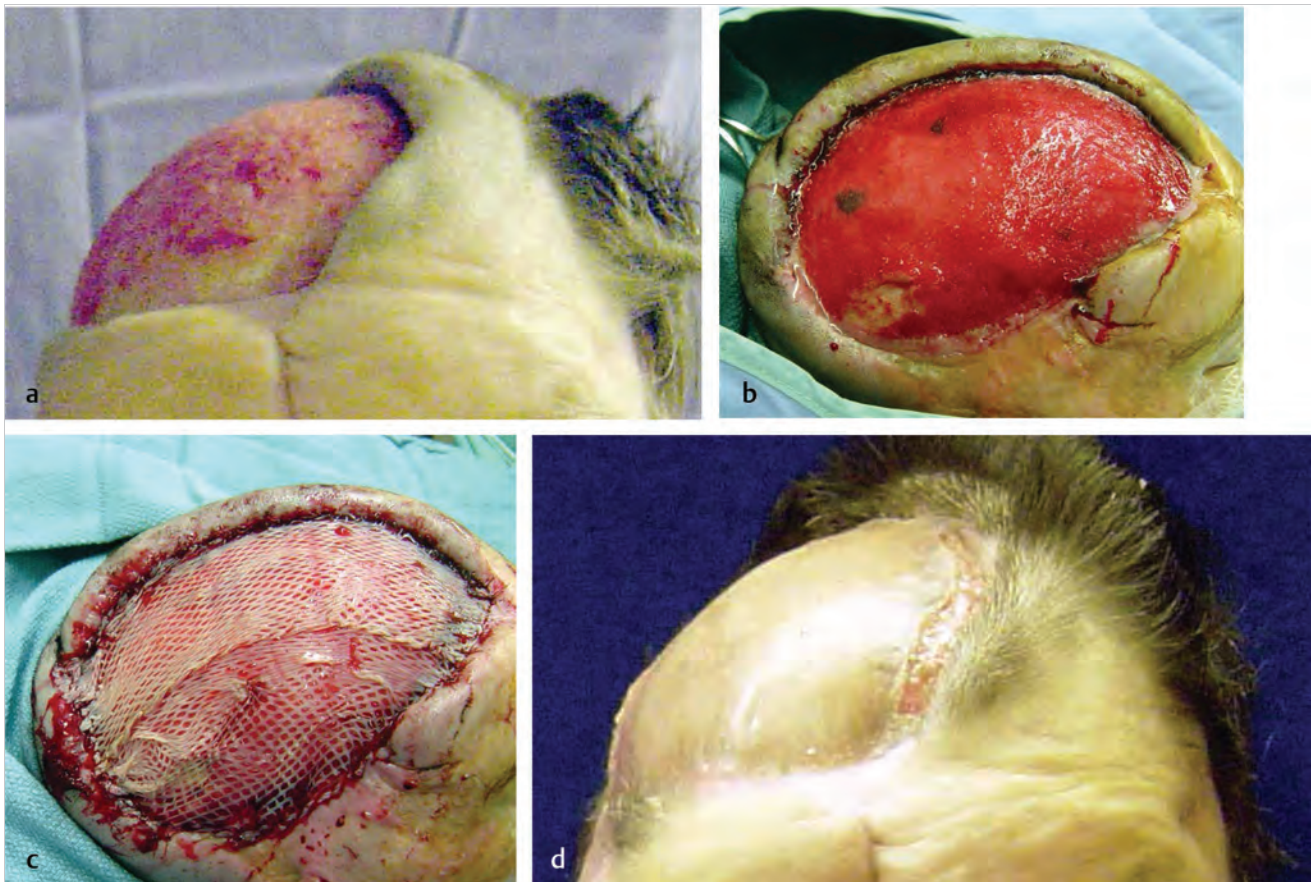


Fig. 24.9 Scalp defect following trauma. (a) Preoperative appearance illustrates absence of periosteum and exposed cranium. (b) Granulation tissue generated after burring of the ectocortex to punctate bleeding from the diploic space followed by negative pressure wound therapy. (c) Split-thickness skin graft placed over granulation tissue. (d) Healed wound amenable to secondary reconstruction with tissue expansion.

24.4.5 Cheek

The most common cheek malformations are associated with hemifacial microsomia (e.g., macrostomia, hypoplasia) and are covered in the chapter devoted to that condition. Other cheek malformations can be associated with facial clefts. The principles of cheek reconstruction for congenital defects are similar, regardless of the type of anomaly. Principles will be exemplified in this chapter by focusing on progressive hemifacial atrophy (also called Parry–Romberg disease). The condition was first noted by Parry (1825) and later described by Romberg (1846).

Hemifacial atrophy is progressive and begins during childhood; both sides of the face are affected equally (► Fig. 24.10). The disease begins at an average age of 9 years and progresses for 3 to 15 years before “burning out.” One-third of patients have only skin and subcutaneous involvement and present at an average age of 15 years. Two-thirds of patients exhibit skeletal deficiency and develop the condition at an average age of 5 years. One-fourth of patients have changes in skin pigmentation and also may have eyebrow alopecia. Patients often exhibit a localized area of atrophy of the forehead, referred to as a *coup de sabre* (“cut of the sword”). Although the etiology of the disease is unknown, it may be a variant of scleroderma. It is hypothesized that a lymphocytic neurovasculitis occurs along the trigeminal nerve.

Reconstruction should not be considered until the disease has completed its progression; intervention usually is started 12 months after the condition has stabilized. Many different techniques have been described to augment the deficient face: adipose grafts, adipose-dermal grafts, dermal substitutes, alloplastic materials, free tissue transfer. Any of these techniques are reasonable, and may be based on the preference of the surgeon. Minor deficiencies are more likely to be amenable to grafting, while major hypoplasia may require free tissue transfer; local–regional flap transfer generally is not an option.

If underlying skeletal hypoplasia is suspected, then computed tomography can help plan reconstruction. Frontal bone, zygoma, maxilla, or mandible deficiency can be enhanced with onlay bone graft or alloplastic material. Patients with orbital deficiency may require floor implants to correct enophthalmos. A severely hypoplastic maxilla or mandible may necessitate an orthognathic procedure after skeletal maturity has occurred (e.g., 16 years in females and 18 years in males).

If a patient requires an incision to remove abnormally pigmented skin, I prefer to use a thick dermal substitute rather than fat grafting because acellular dermis does not resorb and gives a more predictable outcome (► Fig. 24.11). In addition, some reports have shown that fat grafting for progressive hemifacial atrophy has more resorption compared to other recipient sites. If the patient’s occlusion is normal, I augment facial bones



Fig. 24.10 Progressive hemifacial atrophy. (a,b) Preoperative appearance. (c,d) Early postoperative result following reconstruction.

with porous polyethylene (Medpor) rather than bone grafts because there is less resorption and no donor site morbidity. Most patients with hemifacial atrophy can be managed by augmenting the bone and soft tissue with alloplastic material and grafts. Free tissue transfer should be reserved for very severe patients; the scapular and parascapular flaps are most commonly used.

24.4.6 Neck

Pediatric plastic surgeons are consulted to improve soft-tissue deformities of the neck. The most frequent is webbing (also called pterygium colli) from Turner syndrome. Turner syndrome affects 1/3,000 females and results from sex chromosome monosomy (45, X). Although the cause of the web is unknown, it may result from an intrauterine macrocystic lymphatic malformation that expands the skin, deflates prior to birth, and leaves residual tissue and a lowered hairline. The web extends from the mastoid to the clavicle. Other features of Turner syndrome include lymphedema (60%), short stature, amenorrhea,

low-set ears, cardiovascular anomalies (50%), thyroid disorders, renal anomalies, and gonadal dysgenesis.

Several procedures have been described to improve the webbed neck deformity, including Z-plasty, “butterfly” correction, and various flaps. I believe Z-plasties should be avoided because the scars are placed anteriorly and the abnormal hairline is not corrected. The major principles of reconstruction of this defect are (1) remove the abnormally positioned hair on the lower neck, (2) improve neck contour, and (3) position the scars so they are least visible on frontal view. To achieve these goals, I use a posterior neck approach, resection of a large area of abnormal hair and skin, elevation of an anterior skin flap, and repositioning of the skin posteriorly (► Fig. 24.12). The technique is similar to that described by Menick et al in 1984. This procedure hides the incision in the hairline and posterior neck; scars are not visible on front view. If the scars widen on the neck, they can be secondarily narrowed. In my experience, children do not present for correction of their webbed neck until late childhood or early adolescence when their appearance becomes more important to them.



Fig. 24.11 Reconstruction of progressive hemifacial atrophy. The patient underwent (1) excision of atrophied and hyperpigmented skin from the forehead and cheek with linear closure, (2) augmentation of the forehead and cheeks with layered acellular dermis through the areas of skin that were excised, (3) onlay mandible and chin implants with porous polyethylene, (4) improvement of the right upper lip deficiency using a dermal graft harvested from the abdomen, (5) contour of the lower lip by excision of mucosa/submucosa from the left lower lip using a transverse mucosal incision and then using the material as a graft to the right lower lip, and (6) hair transplantation to the right eyebrow. (a) Intraoperative markings. (b) Acellular dermis used to augment the forehead and cheek. (c) Dermal graft inserted into right upper lip. (d) Medpor mandible and chin implants placed on the right side. (e) Occipital scalp used for hair transplantation. (f) Micrografts placed into right medial eyebrow. (g) Forehead “coup de sabre” corrected by resection of the hypoplastic skin, advancement of adjacent forehead, and placement of acellular dermis underneath the area. (h) Intraoperative appearance at the end of the procedure. Bolsters were used to secure acellular dermis under the subcutaneous tissue of the cheek.

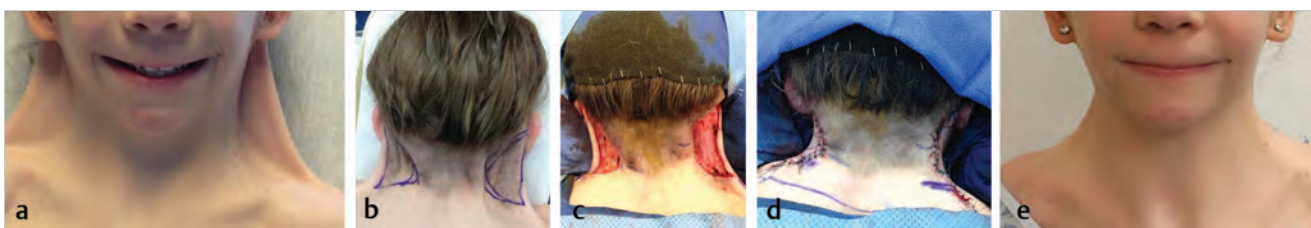


Fig. 24.12 Management of webbed neck. (a) Child with Turner syndrome. (b) Marking of skin excision. (c) Following resection and wide anterior undermining of the anterior skin flap. (d) Posterior inset of anteriorly raised skin flaps. (e) Postoperative appearance.

24.5 Complications

The most frequent unfavorable outcome following reconstruction of scalp and neck deformities is widened scars because of significant tension. Scalp scars can cause visible alopecia, especially in males who have shorter hair. Alopecia can be minimized by maintaining sutures for several weeks, and can be corrected by narrowing the scar secondarily when there is less tension on the wound. After reconstruction of a webbed neck deformity, the scars also can widen because the incision lines

are under significant tension. They can be narrowed secondarily. Wound dehiscence, bleeding, or infection are uncommon.

24.6 Conclusion

Pediatric scalp, cheek, and neck disorders can be reconstructed by many techniques. Generally, procedures as low on the reconstructive ladder as possible should be employed. Because the infant scalp has unique skin laxity, consideration should be

given to extirpation of lesions/alopecia during this period. The major morbidity of these disorders is psychosocial, and thus correcting the problem before 4 years of age often is requested. Progressive hemifacial atrophy must “burn out” before reconstruction. Some families will prefer to wait until the child is older when the patient can participate in the decision to proceed with an operation.

24.7 Key Points

- Cutis aplasia usually can be allowed to heal secondarily; if the dura is exposed, emergent split-thickness skin grafting is necessary.
- Resection of scalp lesions or alopecia is facilitated during the first year of life because of significant skin redundancy.
- Most scalp lesions can be removed with serial excision and do not require tissue expansion.
- Progressive hemifacial atrophy may be improved by several methods; grafts are usually sufficient, but free tissue transfer may be necessary for severe disease.

- Correction of a webbed neck should be performed by skin resection and posterior positioning of anterior skin flaps to avoid scars on frontal view; Z-plasties should be avoided.

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Part IV

Trunk and Lower Extremity

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25 Macromastia

Laura C. Nuzzi and Brian I. Labow

Summary

Macromastia is a common adolescent breast disorder characterized by unilateral or bilateral breast hypertrophy, often resulting in significant functional impairment and negative quality of life. Onset of adolescent macromastia typically occurs shortly after menarche, usually between the ages of 9 and 15, and is distinguished by considerable breast growth out of proportion to the adolescent's frame. The vast majority of patients with macromastia report musculoskeletal pain, painful bra strap grooving, as well as difficulty exercising and participating in sports. Apart from physical symptoms, many patients experience psychosocial distress manifesting in poor mental health, low self-esteem, and increased disordered eating thoughts and behaviors relative to their peers.

Currently, there are no standardized treatment guidelines for adolescents with macromastia. Available non-operative treatments include physical therapy, weight loss and nutrition counsel, hormonal treatments, and ongoing psychological management. However, the efficacy of these treatments is variable, and they often do not completely alleviate symptoms. Macromastia can be successfully treated with early surgical intervention in the form of reduction mammoplasty, though this option is often hindered by various barriers to treatment specific to the adolescent patient. The most widely employed surgical technique in the United States is the Wise pattern resection around an inferiorly based pedicle. Associated musculoskeletal symptoms improve almost immediately following reduction mammoplasty, with breast appearance continuing to improve for several months. Although a relatively safe operation, minor complications related to wound healing in the early postoperative period are common. Conversely, major complications such as tissue loss, loss of sensation, and hemorrhage are very rare.

Keywords: macromastia, breast hypertrophy, adolescent, reduction mammoplasty, breast overgrowth, Wise pattern resection

25.1 Introduction

Macromastia is a common adolescent breast disorder in which the glandular tissue of one or both breasts exhibits hypertrophy (► Fig. 25.1). Onset frequently begins shortly after menarche and is marked by considerable breast growth out of proportion to the adolescent's frame. Aesthetic concerns aside, benign breast overgrowth can result in significant functional impairment and is associated with negative quality of life. The majority of patients present with moderate to severe musculoskeletal pain, bra strap shoulder grooving and bruising, and difficulty when exercising and participating in sports. Adolescents with macromastia are often targets of teasing and harassment and unwanted sexual attention, and as such, many patients suffer from poor mental health and low self-esteem.

Although macromastia can be successfully treated with early surgical intervention, there are several barriers to treatment unique to the adolescent patient. For example, parents, physicians,

and surgeons may be reluctant to offer surgery despite debilitating symptoms, and in some cases insurers set age and weight restrictions for the procedure. Reduction mammoplasty is a well-tolerated procedure with a high satisfaction rate in appropriate patients. In general, patients should be nonsmokers with supportive families and also emotionally and intellectually mature enough to understand the risks and benefits of surgery. Although minor complications related to wound healing are common, major complications such as tissue loss, loss of sensation, and hemorrhage are very rare.

25.2 Diagnosis

Onset of adolescent macromastia occurs shortly after menarche, most commonly between the ages of 9 and 15 years. During this time, the breasts may quickly grow disproportionate to the adolescent's frame and may appear ptotic with striae. In rare cases of virginal breast hypertrophy, rapid and progressive breast development may begin with onset of thelarche and necessitate early surgical intervention. Although breast overgrowth may be noted early by the pediatrician, parent, and patient, the stigmatization of adolescent breast surgery may delay referral of patients with suspected macromastia to the plastic surgeon. By the time the adolescent presents to the plastic surgeon, she may be considerably symptomatic for several years.

Patients with symptoms referable to heavy, ptotic breasts should undergo a complete history, physical examination, and assessment of symptomatology (Box 25.1). A formal "diagnosis" of macromastia can be made using the Schnur Sliding Scale, which examines the relationship between estimated breast



Fig. 25.1 A 17-year-old female with severe macromastia, bra strap grooving, and skin irritation is visible.

resection mass and the patient's calculated body surface area (BSA), in order to determine whether the indications for reduction mammoplasty are primarily medical or cosmetic. A common threshold for insurance coverage is a minimum 500 g of resected tissue per breast, although this may be less in petite or short-statured women.

Box 25.1 Indications for reduction mammoplasty in adolescents

1. Breast symptomatology
 - a) Shoulder grooving and bruising
 - b) Inframammary intertrigo
 - c) Breast ptosis
 - d) Breast striae
2. Associated musculoskeletal pain
 - a) Lower and upper back
 - b) Shoulder
 - c) Neck
 - d) Arm
 - e) Breast
3. Physical impairment
 - a) Daily work, school, life activities
 - b) Exercise and sporting activities
 - c) Secondary scoliosis or postural defects
4. Psychosocial impairment
 - a) Mental health
 - b) Self-esteem
 - c) Disordered eating thoughts and behaviors
 - d) Difficulty finding appropriately fitting clothes and bras
5. Anthropometric characteristics
 - a) Schnur Sliding Scale threshold for BSA

Abbreviation: BSA, body surface area (m²).

Associated musculoskeletal pain—including, but not limited to, the upper and lower back, shoulders, neck, arms, and breasts—should be documented for insurance purposes. Inframammary fold intertrigo, shoulder grooving and bruising from bra straps, and discomfort when exercising should be assessed and noted. During the physical examination, presence of multiple or giant fibroadenoma should be ruled out, particularly in patients presenting with unilateral macromastia or painful, tender breasts.

Adolescents with macromastia may exhibit poorer mental health, lowered self-esteem, and increased disordered eating thoughts and behaviors relative to their peers. Positive exam findings for diminished mental health status, self-esteem, and disordered eating behaviors and obesity should be addressed. The surgeon should also evaluate the patient's skeletal and psychological maturity when considering surgical intervention. The potential for additional breast growth in pubertal patients must be considered and discussed with the patient and parent, but should not absolutely preclude surgery.

25.3 Nonoperative Treatment

Currently, there are no standardized treatment guidelines for adolescent macromastia. Plastic surgeons may delay reduction

mammoplasty due to concerns of additional breast growth and the need for reoperation, patient obesity and potential postoperative complications, and the potential negative psychological effects of breast surgery on adolescents. Age and breast volume cutoffs set by third-party payors may also delay surgery. Patients who are not yet skeletally or emotionally ready for surgery should be followed by the plastic surgeon for exacerbation of symptoms and developmental maturity. Sympathetic reassurance and counsel with respect to future surgical intervention should be provided during these visits. Additionally, if the patient has not been professionally fitted for a bra, she should be encouraged to do so.

The efficacy of estrogen antagonists and steroidal progestins—such as tamoxifen, dydrogesterone, and medroxyprogesterone citrate—to halt breast growth and reduce symptomology is variable. As such, nonoperative treatment should primarily concern minimizing the patient's physical and psychosocial discomfort until surgery can be achieved. Musculoskeletal pain may be temporarily alleviated with referral to physical therapy, and can also be a requirement for reduction mammoplasty coverage by some third-party payors. Weight loss and nutrition counsel may be another requirement for coverage and may offer additional benefits given that adolescent patients with macromastia are more likely to suffer from obesity and disordered eating behaviors.

The psychosocial component of macromastia must not be ignored. Adolescent patients with diminished emotional well-being, self-esteem, and mental health should be referred for ongoing psychological management. During clinic visits, the plastic surgeon should assess current mental status, document worsening of symptoms, and make referrals when necessary. Finding properly fitted bras and clothing also serves as a considerable source of stress for many adolescent macromastia patients. The plastic surgeon may wish to familiarize themselves with specialty clothing and bra outlets that provide greater sizing options and direct patients when necessary.

25.4 Operative Treatment

Reduction mammoplasty is a safe operation requiring a general anesthetic, and may be done on an outpatient basis or a single-night stay. The goal of reduction mammoplasty is to resect excess redundant skin, gland, and adipose tissue of the breast. The breast is reshaped and nipple areola complex (NAC) is repositioned and resized to provide proper position and contour. Care must be taken to preserve breast mound projection when repositioning and resizing the length of the inframammary fold length and NAC diameter. Nipple placement should be at or slightly above the inframammary fold, and is crucial in maintaining aesthetic and three-dimensional appearance of the breast.

Reduction mammoplasty can be performed using a variety of pedicles and incision patterns. The most widely employed technique in the United States is the Wise pattern resection around an inferiorly based pedicle. The Wise pattern is versatile and particularly advantageous for large reductions, ptotic breasts, patients with poor skin quality, and in instances requiring considerable nipple repositioning. This

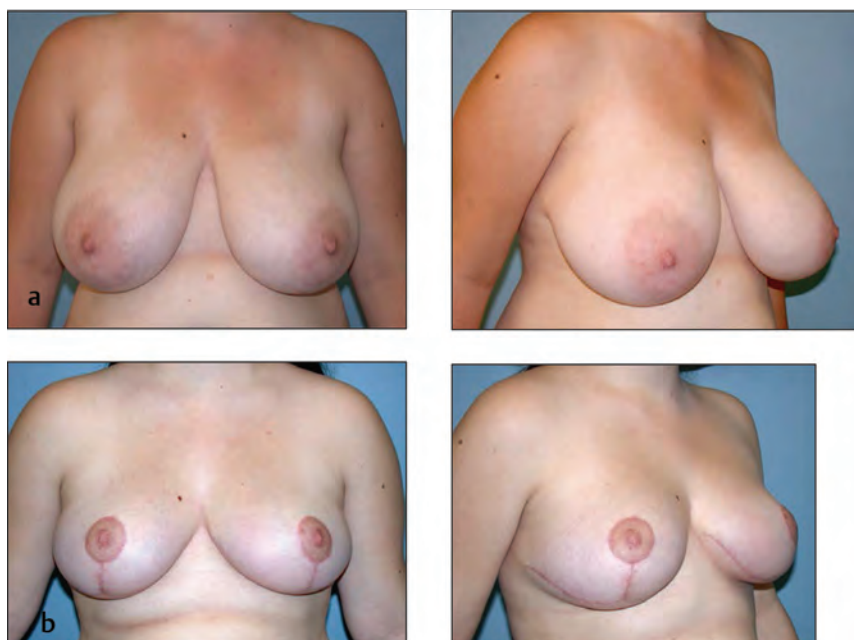


Fig. 25.2 (a) A 16-year-old with moderate bilateral adolescent macromastia, DD cup-size. (b) Nine months following Wise pattern reduction mammoplasty, C cup-size, 750 total grams removed.



Fig. 25.3 (a) An 18-year-old with severe bilateral adolescent macromastia, H cup-size. (b) Two and half years following reduction mammoplasty, D cup-size, 4,530 total grams removed.

technique is most likely to leave sensation and ability to lactate intact; however, some feel the potential for visible scarring and limited breast projection is greater using the Wise pattern (► Fig. 25.2, ► Fig. 25.3).

25.4.1 Postoperative Management

Postoperative routines vary by surgeon. However, following closure, the surgical site is often covered with steri-strips, absorbent dressings, and a surgical support bra. At the first postoperative visit, the dressing is removed and the surgical site checked. Typically, patients require 3 to 5 days of prescription pain medication and are out of school or work for

roughly 1 week. Exercise and lifting should be avoided for the first 6 weeks following surgery, after which full activity can be resumed. Similarly, patients should use a supportive sports bra and avoid a bra with an underwire for 6 weeks following reduction mammoplasty.

Associated musculoskeletal symptoms improve almost immediately following reduction mammoplasty, with breast appearance continuing to improve for several months. Lateral breast sensation normalizes around the sixth month mark; however, scar maturation may take 1 to 2 years to achieve optimal appearance. In this population, it may be advantageous to follow patients at least several times during the first postoperative year and annually thereafter.

25.5 Complications

Although reduction mammoplasty is a relatively safe and common procedure, roughly 40 to 50% of patients will experience some complication. Despite these complications, patient satisfaction following reduction mammoplasty remains high. Early postoperative complications are more frequent, but usually minor. Wound dehiscence and delayed wound healing present in 10% of patients. Additional clinic visits may be warranted for wound dehiscence worrisome to the patient and can be managed with dressing changes. Exposed sutures causing pain and minor skin irritation from steri-strips are also common. Rates of infection requiring oral antibiotics and drainage are less than 1%. Excessive bleeding, seroma, and hematoma requiring reoperation and evacuation are rare. Risks of nipple ischemia and fat necrosis are exceedingly low in the adolescent population.

Patients should be followed closely during the first postoperative year and beyond to assess longer term complications. Diminished nipple and lateral breast sensation is to be expected during the early healing period. Patients should regain sensation within the first 6 months; however, a subset of women will have persistent partial or complete loss of breast and/or nipple sensation following reduction mammoplasty.

Keloid and hypertrophic scar formation may be noted after several months of wound healing. Scar massage, silicon sheeting, and eventually corticosteroid injections can be employed in the clinic setting to decrease the contour of existing keloids and to diminish burning and pruritic symptoms.

A considerable number of adolescents present preoperatively with mild to moderate breast asymmetry in addition to macromastia, and in fact a degree of NAC or breast asymmetry is to be expected postoperatively. As the breast skin relaxes and glandular tissue settles after surgery, the breast may appear to “bottom out.” This results in a breast where the nipple appears to rest too high above the inframammary fold. In general, it is best to ensure that the nipple is not positioned too high at the time of surgery. This is especially true in patients with thin skin. Revisional procedures can correct size and position asymmetries of the breast and NAC, but these should be considered after the first postoperative year. Dog-ears, or redundant skin at the suture sites, can be addressed through limited revisions under local anesthesia.

Unique to the adolescent patient is the risk of reoperation due to continued pubertal or future gestational breast growth. Although this can occur, age alone should not be viewed as an absolute contraindication to surgery. For many patients, the physical and psychosocial benefits of early surgical intervention may far outweigh the risk of a repeat procedure later in life. It should be noted, however, patients with a 1-year history of stable breast size typically do not exhibit considerable breast regrowth following reduction mammoplasty. An additional concern of referring pediatricians, plastic surgeons, and patients alike is the effect of reduction mammoplasty on an adolescent's ability to lactate and produce sufficient milk volume in the future. Postoperative lactation ability may be associated with surgical technique. All efforts should be made to spare the neurovascular structures of the ductal breast tissue. Postoperative patients are believed to have a 65% lactation success rate; however, many patients may require formula supplementation.

25.6 Conclusion

Adolescent macromastia is a common hyperplastic breast disorder characterized by breast overgrowth disproportionate to the patient's frame. Patients may be highly symptomatic and present with musculoskeletal pain, breast striae, and painful shoulder grooving and bruising from bra straps. Macromastia can result in significant functional and psychosocial impairment and is associated with negative health-related quality of life. However, the adolescent patient faces several unique barriers to treatment that may delay referral and surgical intervention. Age and potential for breast regrowth and reoperation should not preclude surgery in this otherwise healthy patient population.

Early surgical intervention may alleviate and prevent the exacerbation of physical and psychosocial symptoms into adulthood. In many cases, this resolution of symptoms may outweigh the risk of reoperation later in life. Reduction mammoplasty in the adolescent is a well-tolerated procedure with a high rate of satisfaction. Postoperative complications are common, but typically minor and can be largely managed in the clinic setting.

25.7 Key Points

- Macromastia is a common hyperplastic adolescent breast disorder resulting in considerable unilateral or bilateral breast overgrowth.
- Patients may suffer from functional and psychosocial impairments due to their breast size. Macromastia is highly associated with moderate to severe musculoskeletal pain.
- Reduction mammoplasty is the most effective treatment for macromastia in appropriately selected patients.
- Age and potential for additional growth should not preclude surgery; early intervention can prevent the exacerbation of physical and psychosocial symptoms in adulthood.
- Postoperative complications are common, but minor, and can be largely managed in the clinic setting.

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26 Gynecomastia

Laura C. Nuzzi and Brian I. Labow

Summary

Gynecomastia is a common finding among healthy adolescent boys. Benign excess glandular breast tissue is the hallmark of gynecomastia. Severity ranges from a puffy appearing nipple areolar complex (NAC) to marked enlargement and ptosis. Gynecomastia may result in significant distress, and diminished self-esteem and mental health.

A comprehensive history and physical examination underpin the assessment of gynecomastia; these may be supplemented by laboratory studies and imaging. A thorough history must include a medications history, an inquiry into recreational and performance enhancing drugs, an exploration of symptoms of endocrine disease, and a systems review. Breast palpation is crucial and should aim to determine the degree of glandular and adipose involvement, and the presence of pain, tenderness and breast masses.

The majority of cases are idiopathic and self-resolving; these patients are managed with reassurance and follow-up until resolution. Other patients experience gynecomastia secondary to pathological processes or pharmacological side effects, which may be amenable to medical intervention. Non-operative interventions include the use of compression shirts, nutritional management, medication review, and referral to endocrine services for specialized treatment. Surgical intervention may be necessary in response to gynecomastia that is severe, distressing, or persistent. Operative treatment may be a combination of mastectomy, mastopexy, liposuction, and lipectomy. Satisfaction with surgery remains high among patients despite relatively frequent minor post operative complications.

Keywords: gynecomastia, breast, mastectomy, liposuction, lipectomy

26.1 Introduction

Pubertal gynecomastia is a common finding among otherwise healthy adolescent boys. The condition is marked by benign excess glandular breast tissue, although all breast mounds contain gland, fat, and skin to a variable degree. Severity encompasses a broad spectrum of presentations ranging from a puffy appearing nipple areolar complex (NAC) to marked breast enlargement and ptosis. Adolescents with gynecomastia of any severity may suffer from poor mental health and low self-esteem. It should be noted that most boys will exhibit some degree of gynecomastia during adolescence, with the majority of cases self-resolving 2 to 3 years after onset. Although pubertal gynecomastia is largely idiopathic, proper

physical examination and workup is essential to rule out potential underlying pathological causes.

Most idiopathic cases of mild gynecomastia can be managed with sympathetic reassurance and yearly follow-up until resolution. The efficacy of medical intervention is variable and dependent on etiology. Persistent or distressing gynecomastia may warrant surgical intervention, with technique determined by the extent of hypertrophy and skin excess. The goal of treatment is to reduce the projection of the breast mound and NAC. Excision of redundant skin and reduction of NAC size may also be performed in cases of moderate to severe gynecomastia. Surgical complications are more common, but typically minor in patients with severe gynecomastia who undergo larger tissue resections.

26.2 Diagnosis

26.2.1 Presentation

Gynecomastia is a common finding among adolescent boys, particularly during Tanner stages 2 and 3. It is estimated that by the age of 14 years, 70% of all adolescent boys will exhibit some degree of gynecomastia. The majority of cases are idiopathic and self-resolving, with over 90% of affected boys showing complete resolution of symptoms within 3 years of onset. As a result, gynecomastia is often managed in the pediatric primary care setting. Referral to the plastic surgeon, however, may be necessary in cases of distressing, persistent, or considerable gynecomastia.

Male breast hypertrophy can be familial and may present unilaterally or with some degree of asymmetry in up to one-third of patients. Unlike macromastia, formal “diagnosis” of gynecomastia does not require meeting a threshold mass of tissue resected. The degree of breast hypertrophy in gynecomastia may instead span a range of mild to severe. As such, gynecomastia is described using a graded scale often used by third-party payors to determine insurance coverage eligibility (Box 26.1; ► Fig. 26.1). Grade I gynecomastia is the mildest form, with minimal hypertrophy (<250 g) without ptosis. Grade II gynecomastia is defined by moderate hypertrophy (250–500 g) without ptosis. Grade III gynecomastia (>500 g) describes severe hypertrophy with skin excess, and is marked by mild ptosis. Grade IV gynecomastia (>500 g) is the most severe with considerable hypertrophy, skin excess, and moderate to severe ptosis. It should be noted that gynecomastia of any grade may result in significant distress and diminished self-esteem and mental health.



Fig. 26.1 Graded severity of gynecomastia. (a,b) Grade I: mild breast hypertrophy without ptosis. (c,d) Grade II: moderate breast hypertrophy without ptosis. (e,f) Grade III: moderate breast hypertrophy with mild ptosis and skin excess. (g, h) Grade IV: severe breast hypertrophy with considerable ptosis and skin excess.

Box 26.1 Graded Severity of Gynecomastia

- Grade I
 - <250 g hypertrophy excess per side
 - Minimal glandular, adipose involvement
 - No skin involvement or ptosis
 - Grade Ia: breast mound composition primarily adipose tissue
 - Grade Ib: breast mound composition primarily glandular tissue
- Grade II
 - 250–500 g hypertrophy excess per side
 - Moderate glandular, adipose involvement
 - No skin involvement
 - No ptosis
 - Grade IIa: breast mound composition primarily adipose tissue
 - Grade IIb: breast mound composition primarily glandular tissue
- Grade III
 - >500 g hypertrophy excess per side
 - Moderate to severe glandular, adipose involvement
 - Mild to moderate skin involvement
 - Mild ptosis (grade 1)
- Grade IV
 - >500 g hypertrophy excess per side
 - Severe glandular, adipose involvement
 - Moderate to severe skin involvement
 - Moderate to severe ptosis (grade II or III)

Source: Adapted from Rohrich RJ, Ha RY, Kenkel JM, Adams WP Jr. Classification and management of gynecomastia: defining the role of ultrasound-assisted liposuction. *Plast Reconstr Surg* 2003;111:909–923.

True gynecomastia is a proliferation of male glandular breast tissue, and should be distinguished from pseudogynecomastia in which the majority of the breast mound is secondary to excess adipose tissue. Both tissue types, however, are present to varying degrees in patients with true pubertal gynecomastia. Adolescent gynecomastia is highly associated with obesity. Excess peripheral adipose tissue can contribute directly to the size of the breast mound. Additionally, the presence of elevated aromatase in adipose tissue can result in glandular overgrowth by stimulating the conversion of androgens to estrogen. Regardless, it should be emphasized that it is usually the size and contour of the breast mound rather than its relative composition that is concerning to patients.

26.2.2 Physical Exam and Workup

All patients presenting to the plastic surgeon with suspected gynecomastia should undergo a complete history and physical exam. A breast exam should be performed to determine the degree of glandular and adipose tissue involvement and to assess for pain, tenderness, and palpable breast masses. Patients are most likely to experience pain and tenderness during the first 6 months of symptoms when the breast mound is undergoing ductal proliferation. Presence of glandular involvement

can be easily palpated on exam as a rubbery, mobile disc lying beneath the NAC. The majority of adolescents presenting with gynecomastia are otherwise healthy. Pubertal physiologic gynecomastia absent of any risk factors or findings on physical examination can be followed without additional studies.

Prescription and illicit drug use should be assessed. Drug-induced adolescent gynecomastia is commonly associated with excessive marijuana and alcohol use, and to a lesser extent, H₂ receptor antagonists, psychoactive agents, cardiovascular drugs, antiandrogens, chemotherapeutics, antibiotics, and antiretrovirals. The use of anabolic steroids and over-the-counter nutraceuticals to improve strength or athletic performance should also be discussed.

Additional studies should be performed for patients with persistent or progressive gynecomastia, breast hypertrophy arising during late puberty, palpable testicular or breast masses, hypogonadism, or delayed puberty. Laboratory studies may include free testosterone (T), luteinizing hormone, follicle-stimulating hormone, estradiol, and beta-human chorionic gonadotrophin tests. Low testosterone with or without elevated gonadotrophin levels may be indicative of Klinefelter syndrome (46, XXY). A karyotype should be obtained in cases where testicular atrophy or lab values are suggestive of hypogonadism. In rare suspected cases of renal, hepatic, or thyroid disease, liver function tests and calcium, creatinine, and thyroid-stimulating hormone assays may be necessary.

Imaging of the breast is rarely performed except when a suspicious mass is noted on exam or if required to document glandular excess. Of note, adolescent male breast cancer is extremely rare and idiopathic gynecomastia is not associated with an elevated risk.

26.3 Nonoperative Treatment

As gynecomastia is largely idiopathic and self-resolving, the majority of cases may be managed with support and sympathetic reassurance. These patients should be monitored periodically every 6 to 12 months until resolution. Gynecomastia is strongly associated with poor psychological well-being and self-esteem, and the plastic surgeon should use clinic visits to monitor mental health, document changes, and make referral for psychological management when necessary.

Providers may recommend the use of compression shirts worn under clothing as an immediate, inexpensive nonmedical, and nonsurgical means to minimize the contour of the chest and provide support. This may be especially beneficial for patients who report teasing, bullying, or poor self-esteem as a result of their chest size.

Gynecomastia is associated with obesity, and treatment should include weight loss and nutritional management. Patients with moderate to severe glandular and skin involvement should be advised that although weight loss may be beneficial for a myriad of health reasons, weight loss alone may not be sufficient to address the problem. Weight loss, however, may reduce chest size for patients with pseudo-gynecomastia in which the majority of the breast mound is composed of adipose tissue. These patients may wish to seek surgical intervention following weight loss for excision of any remaining glandular tissue and redundant skin. A reduction of trunk adipose during early puberty, closer to the onset of presentation, may abrogate

glandular hypertrophy caused by paracrine signaling. As such, younger obese patients should be encouraged to lose weight as a means to curtail future breast growth.

Gynecomastia secondary to drug use should be managed by discontinuing the use of illicit or performance-enhancing drugs, and counsel with the prescribing physician in cases of prescription medication-induced overgrowth. Gynecomastia persisting for 12 months following changes in drug use should receive additional workup and counsel with respect to surgical intervention.

Patients with suspected hormonal causes should be referred to an endocrinologist with experience in treating the adolescent patient. Medical treatment for endocrine-related gynecomastia may include anti-estrogens or aromatase inhibitors, with androgen use falling out of favor. Efficacy is highly variable and patients should be advised that pharmacotherapy is most effective during early onset before fibrous tissue replaces gland. Surgical intervention should be discussed in cases of distressing gynecomastia secondary to underlying medical cause when satisfactory improvement cannot be achieved with pharmacotherapy alone.

26.4 Operative Treatment

Surgical intervention may be warranted in cases of persistent gynecomastia resulting in considerable emotional distress. Some providers may wish to delay surgery until skeletal or sexual maturation is complete due to possibility of additional growth and potential need for reoperation. However, the benefits of early surgical intervention may offset the risk of reoperation, particularly in patients with severe gynecomastia or those suffering from teasing, bullying, and considerable psychological impairment due to the appearance of their chest. Early surgical intervention may alleviate and prevent the exacerbation of psychosocial symptoms into adulthood.

Management is highly dependent on the grade of gynecomastia and the degree of glandular, adipose, and skin involvement. Mild to moderate gynecomastia (grades I–II) can be treated using less invasive techniques. In these instances, a sufficient reduction in volume and contour can be achieved using a combination of simple excision through a periareolar incision, ultrasound-assisted liposuction, or standard suction-assisted lipectomy to reduce the potential for scarring (► Fig. 26.2). Extremely mild cases limited to the NAC may require only simple excision and can be performed in the clinic. In cases marked predominately by excessive glandular tissue (grades Ib and IIb), some have reported that ultrasound-assisted liposuction may be advantageous over standard suction-assisted lipectomy. At lower energy settings, ultrasound-assisted liposuction effectively removes adipose tissue while sparing higher density structures; however, higher settings can target dense fibroglandular tissue.

A combination of subcutaneous mastectomy to remove gland using a periareolar incision, mastopexy to elevate and resize the NAC, and liposuction is employed in moderate to severe cases of gynecomastia (► Fig. 26.3). Most commonly, either a concentric periareolar approach or an inferior pedicle and inframammary or Wise- pattern incision are used. In some severe cases, a complete mastectomy with free-nipple grafting can also be used. A compression vest and drains may be used for a short postoperative period. Mild to moderate discomfort may persist for up to 5 days following surgery and can be managed using over-the-counter and prescription analgesics. The patient may remove dressings and shower after the first office visit, leaving the steri-strips in place to self-detach. Patients should stay out of school or work for roughly 1 week, and exercise and heavy lifting should be avoided for the first 4 to 6 weeks following surgery, after which full activity can be resumed.

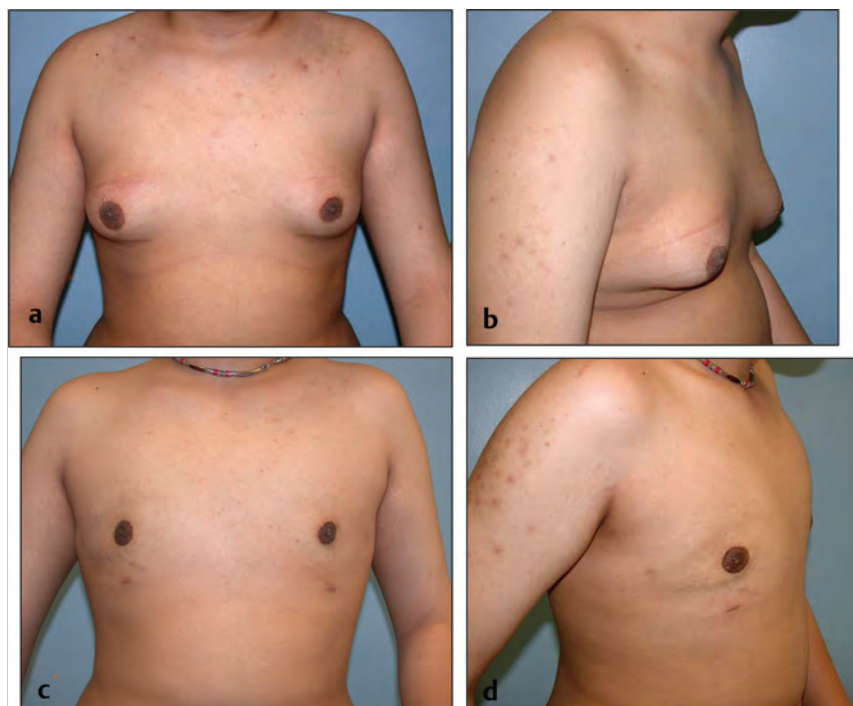


Fig. 26.2 (a,b) A 16-year-old with Grade II adolescent gynecomastia. (c,d) One year following suction lipectomy, and transareolar simple mastectomies.



Fig. 26.3 (a,b) A 17-year-old with Grade III adolescent gynecomastia. (c,d) Three years following suction lipectomy, bilateral simple mastectomies, and concentric mastopexies.

26.5 Complications

Complication type and severity following surgical correction of gynecomastia largely depend on treatment type. Patients undergoing liposuction for grade I gynecomastia are more likely to have fewer and less serious complications than a patient undergoing mastectomy for grade IV gynecomastia. Perioperative complications include those inherent in most procedures performed in the operating room. Complications are typically less prevalent and serious when surgical intervention is performed by a plastic surgeon.

Early postoperative complications are typically minor and more common in large volume resections. The most common early complications are seroma and hematoma, which can be managed with evacuation and drains as needed. Postoperative infection is exceedingly rare in periareolar incisions and can be treated using oral antibiotics.

Keloid and hypertrophic scar formation may occur, particularly in patients undergoing large resections via a long inframammary incision. Following scar maturation, scar revisions can be performed in the clinic or operating room. Corticosteroid injections can be employed in the clinic setting to decrease the contour of existing keloids, and minimize pain and pruritic symptoms.

Long-term complications may include asymmetry and contour irregularities, particularly following liposuction, lipectomy, and large resections. An under-resection of peripheral tissue can result in considerable deformation, while an over-resection of tissue near the NAC can result in insufficient projection and a saucerlike deformity. Loose skin is to be expected following large resections and may improve over the first year in patients who are young, still growing, and have good skin quality. Patients should be advised during the preoperative visit that persistent lax skin may require correction in the future or

additional skin resection at the time of the primary operation. Breast regrowth following surgical correction is unlikely to occur in older patients or those with moderate to severe gynecomastia. However, younger patients who have not completed puberty or patients with gynecomastia of an uncontrolled, pathological origin may exhibit additional growth of any remaining glandular tissue following surgery.

26.6 Conclusion

Pubertal gynecomastia is a common finding among otherwise healthy adolescent boys. True gynecomastia is marked by the proliferation of glandular breast tissue of varying severity, although excess adipose tissue is commonly present as well. Most cases of idiopathic gynecomastia are self-resolving and require only sympathetic reassurance. Proper workup is essential in suspected drug-induced, or pathological, cases of gynecomastia. Regardless of composition, cause, or severity, gynecomastia can result in significant mental health and self-esteem deficits. During clinic visits, the plastic surgeon should assess current mental status and make referrals when necessary.

Surgical intervention may be warranted in cases of persistent, severe, or distressing gynecomastia. Early surgical intervention may alleviate and prevent the exacerbation of psychosocial symptoms into adulthood. Surgical treatment is largely dependent on the severity and grade of gynecomastia. Mild gynecomastia of grades I or II may be managed using simple excision or liposuction performed in either the clinic or the operating room. Treatment of moderate to severe gynecomastia is more invasive and may necessitate the need for larger composite resections. Complications, though typically minor, are common and are more frequently associated with larger resections.

26.7 Key Points

- Gynecomastia is the benign overgrowth of glandular breast tissue in males. Breast hypertrophy is a common finding in healthy adolescent boys and is mostly idiopathic and self-resolving.
- Patients may suffer from psychological and self-esteem deficits due to their chest size.
- Gynecomastia is described using a graded scale of I to IV, encompassing varying degrees of severity and glandular, adipose, and skin involvement.
- A combination of mastectomy, mastopexy, liposuction, and lipectomy can be employed to achieve a sufficient reduction in the size and contour of the breast mound while resizing and repositioning the nipple areolar complex nipple areolar complex (NAC).
- Satisfaction with surgery remains high among patients despite relatively frequent minor postoperative complications.
- Revision surgeries may be performed in instances of persistent skin laxity, hypertrophic scar formation, and postoperative asymmetry.

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27 Breast Asymmetry

Laura C. Nuzzi and Brian I. Labow

Summary

Breast asymmetry is not a formal diagnosis, but rather a term that encompasses normal and abnormal variances in breast size, shape, or position of idiopathic, congenital, or acquired origin. Although breast asymmetry is exceedingly common during adolescence, abnormal asymmetry that persists into late adolescence and early adulthood has the potential to negatively impact an adolescent's quality of life, psychosocial well-being, and self-esteem. A variety of hyperplastic and hypoplastic breast conditions can result in asymmetry. Additionally, adolescents may present with acquired breast asymmetry secondary to benign masses, prior breast surgery, or trauma to the breast bud or gland.

Breast asymmetry can be managed by non-surgical or surgical means depending on the severity of the difference and level of patient distress. As some degree of asymmetry is common during thelarche, it is generally advisable to observe younger adolescent patients until they are close to completing or have completed skeletal growth. Breast prostheses or bra inserts may be of particular benefit to younger patients who are distressed by the uneven contour and size of their breasts given these devices immediately improve appearance, can be adjusted easily with growth, and can be worn during exercise and under swimsuits. Abnormal, persistent, or psychologically distressing asymmetry can be surgically corrected once the patient reaches skeletal and emotional maturity. The plastic surgeon may employ a combination of breast augmentation, mastopexy, and reduction mammoplasty to improve symmetry and meet the individual goals of the patient. Postoperative complications vary by the type of surgical operation performed, though the potential need for revisional procedures should be emphasized preoperatively for this population.

Keywords: breast asymmetry, adolescent, breast augmentation, mastopexy, reduction mammoplasty, abnormal asymmetry

27.1 Introduction

Breast asymmetry is exceedingly common during adolescence. Thelarche is marked by glandular proliferation of varying rates and may result in uneven-appearing breasts early in development. Pubertal breast asymmetry typically improves or completely resolves as breast growth slows during late adolescence. Mild breast differences, however, may persist into adulthood and is a normal finding on examination. Breast asymmetry is not a formal diagnosis, but a catch-all term encompassing a spectrum of normal and abnormal breast differences of various etiologies (► Fig. 27.1; Box 27.1).

Box 27.1 Etiology of Adolescent Breast Asymmetry

1. Pubertal
2. Hyperplastic breast condition
 - a) Macromastia
 - b) Virginal breast hypertrophy
3. Hypoplastic breast condition
 - a) Amazia
 - b) Amastia
 - c) Breast hypoplasia
 - d) Poland syndrome
 - e) Tuberous breast deformity
4. Acquired
 - a) Benign palpable masses
 - b) Malignant neoplasms
 - c) Prior breast surgery
 - d) Trauma

The burden of adolescent breast asymmetry is primarily psychological. Mild to severe breast asymmetry can negatively impact an adolescent's quality of life, psychosocial well-being, and self-esteem. Patients may report difficulty finding bras that properly fit each breast, embarrassment or teasing among peers, and reluctance to wear fitted clothing and swimsuits that highlight breast irregularities.

Nonsurgical management aims to immediately improve cosmesis and self-esteem through the use of prosthetic bra inserts and custom bras. Surgery may be warranted in cases of distressing or abnormal asymmetry that persists into late adolescence or early adulthood. The type of surgical intervention is dictated by the individual patient's breast differences and expectations. The plastic surgeon may employ a combination of breast augmentation, mastopexy, and reduction mammoplasty to achieve symmetry and meet the individual goals of the patient.

27.2 Diagnosis

Breast asymmetry is not a formal diagnosis, but rather an umbrella term describing normal and abnormal differences in breast size, shape, or position of idiopathic, congenital, or acquired origin. Presentation and degree of severity can vary widely; however, most adolescents with breast asymmetry will exhibit normally developed breasts of unequal sizes, ranging from less than one cup size difference between breasts to that of four or more cup sizes. Asymmetry is common, and an estimated 90% of all women will experience some degree of breast



Fig. 27.1 Asymmetry of varying severity and etiology. (a,b) A 16-year-old with mild to moderate breast asymmetry (250 mL, or two-cup-size difference). (c,d) A 19-year-old with severe breast asymmetry (1,250 mL, or four-cup-size difference). (e,f) A 17-year-old with breast asymmetry secondary to giant fibroadenoma excision 4 years prior (arrow points to well-healed incision). (Continued)



Fig. 27.1 (Continued) (g) A 15-year-old with Poland syndrome, right-sided amazia, and ipsilateral symbrachydactyly.



Fig. 27.2 A 15-year-old patient with mild asymmetry and severe tuberous breast deformity.

asymmetry over their lifetime. Most adolescent cases are idiopathic and the byproduct of normal glandular proliferation during breast development. Onset of pubertal asymmetry is common at thelarche (Tanner stage 2) and improves over the course of breast maturation.

A variety of hyperplastic and hypoplastic breast conditions may yield asymmetries in volume, shape, and position. Unilateral macromastia or asymmetric bilateral macromastia will result in one breast that is considerably hypertrophic and ptotic relative to the contralateral breast. These patients may present with musculoskeletal pain, bra strap grooving, breast striae, and inframammary fold intertrigo. Formal “diagnosis” of unilateral or asymmetric macromastia can be supported using the Schnur Sliding Scale. Many insurers utilize this scale, which uses calculated body surface area to determine the breast resection mass necessary for a reduction mammoplasty to be considered medically indicated.

Breast asymmetry secondary to a hypoplastic breast condition is less common and presents with varying degrees of unilateral or bilateral breast insufficiency. Breast hypoplasia and amazia are defined, respectively, by an insufficiency or complete absence of breast tissue with intact pectoralis major muscle and nipple areolar complex (NAC). Amastia, however, is an extremely rare congenital anomaly marked by the absence of both the breast gland and the NAC. Tuberous breast deformity is a developmental anomaly resulting in a constricted breast, a high inframammary fold, and pseudo-herniation of the breast

gland through the areola (► Fig. 27.2). Ptosis, breast hypoplasia, and skin insufficiency typically accompany moderate to severe tuberous breast deformity. Poland syndrome is a rare congenital chest anomaly affecting both males and females. Patients will present with partial or complete unilateral absence of the pectoralis major muscle, and commonly ipsilateral upper extremity anomalies. Females may also exhibit bilateral or unilateral breast hypoplasia or amazia along with a superiorly displaced and hypoplastic NAC. The majority of patients with Poland syndrome are diagnosed at birth, although extremely subtle cases may not be diagnosed until adolescence.

Adolescents may also present with acquired breast asymmetry secondary to benign masses (e.g., fibroadenoma, hemangioma, and vascular malformation), prior breast surgery, or trauma to the breast bud or gland. Presence of fibroadenoma should be ruled out in patients presenting with breast pain or tenderness.

27.3 Nonoperative Management

As some degree of asymmetry is common during thelarche, it is generally advisable to observe younger adolescent patients over time for resolution or worsening of symptoms. Adolescent asymmetry is largely idiopathic with considerable improvement typically appreciated by Tanner Stage 4. Patients should be monitored at least annually, and sympathetic reassurance and counsel with respect to management should be provided during these visits.

The psychosocial impact of breast asymmetry must not be ignored. Adolescents with varied diagnoses and degrees of severity may suffer diminished emotional well-being, self-esteem, and social functioning. Patients may report teasing or embarrassment among peers due to the appearance of their chest. During clinic visits, the plastic surgeon should assess current mental status, document worsening of symptoms, and make referral for psychological management when necessary. As breast asymmetry is associated with obesity, weight loss and nutritional management should also be encouraged. Patients should be advised, however, that weight loss alone will not alleviate the appearance of their breast asymmetry, particularly in cases of unilateral or asymmetric macromastia with substantial glandular involvement.

Although surgery should be deferred until skeletal and emotional maturity is reached, nonsurgical intervention may help offset the negative psychosocial impact of breast asymmetry. Finding properly fitted bras and clothing may serve as a source of stress for adolescents with asymmetry. The plastic surgeon may wish to become familiarized with clothing stores that provide seamstress services to join bra cups of differing sizes to create a single garment that properly fits each breast. Breast prostheses or bra inserts may be of particular benefit to adolescent patients who are distressed by the uneven contour and size of their breasts. These devices immediately improve cosmesis and many can be worn during exercise and under swimsuits.

27.4 Operative Treatment

Abnormal, persistent, or psychologically distressing asymmetry can be surgically corrected once the patient is skeletally and emotionally mature. The majority of patients seeking surgical

intervention for idiopathic asymmetry or breast hypoplasia opt for breast augmentation. Improved symmetry with the contralateral breast can be achieved through unilateral or bilateral implant placement, with or without mastopexy, and is largely dependent on the individual needs of the patient.

Most patients with breast asymmetry secondary to a developmental or congenital anomaly will undergo surgery to correct deformations in breast shape and position to restore symmetry. Severe hypoplasia, amazia, or Poland syndrome may require a staged approach using tissue expansion to increase the skin envelope prior to definitive implant placement.

Correction of tuberous breast deformity aims to release the constricted base of the breast, lower the inframammary fold, correct the herniated appearance of the NAC, and increase breast volume and skin sufficiency if necessary. This typically requires a periareolar approach with removal of a concentric ring of excess areolar skin, subglandular scoring, lowering the inframammary fold, and a subglandular or subpectoral breast implant (► Fig. 27.3). Treatment for severe tuberous breast deformity may require tissue expansion prior to definitive treatment. Mastopexy may be needed in cases of moderate to severe ptosis to reposition the NAC above the inframammary fold. Treatment of Poland syndrome requires a more complex reconstruction, typically employing a latissimus dorsi transfer transposed over the right chest wall to support the placement of tissue expander or breast implant. Although implant-only reconstructions can be used in mild cases, the results are generally less satisfying in moderate to severe cases. Autologous free-tissue transfer can also be performed in cases where there is a larger-sized contralateral breast or when the patient prefers to avoid an implant. If this is the case, it is important to ensure the presence of adequate recipient vessels preoperatively and establish a backup plan

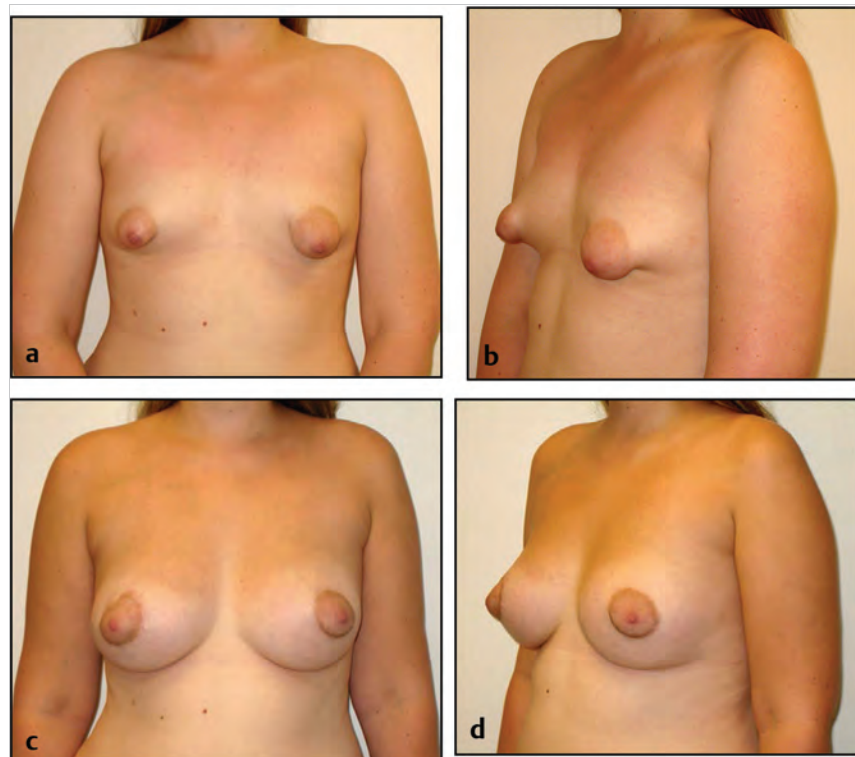


Fig. 27.3 A 19-year-old with tuberous breast deformity (a,b) preoperatively and (c,d) 1 year following correction with areolar size reduction, subglandular scoring, and subpectoral 325 mL, smooth, round, saline implants.

in case they are hypoplastic or absent. Contralateral mastopexy may be performed to further improve symmetry (► Fig. 27.4).

In instances of unilateral macromastia or asymmetric bilateral macromastia, either unilateral reduction mammoplasty or bilateral asymmetric reduction can be performed (► Fig. 27.5).

Estimating volume differences preoperatively can be facilitated using sizers and appropriate full-coverage unpadded bras. In some instances, a reduction on the larger side and a mastopexy alone on the contralateral side may be sufficient. A variety of techniques exist for all of these options and all work well (► Fig. 27.6).

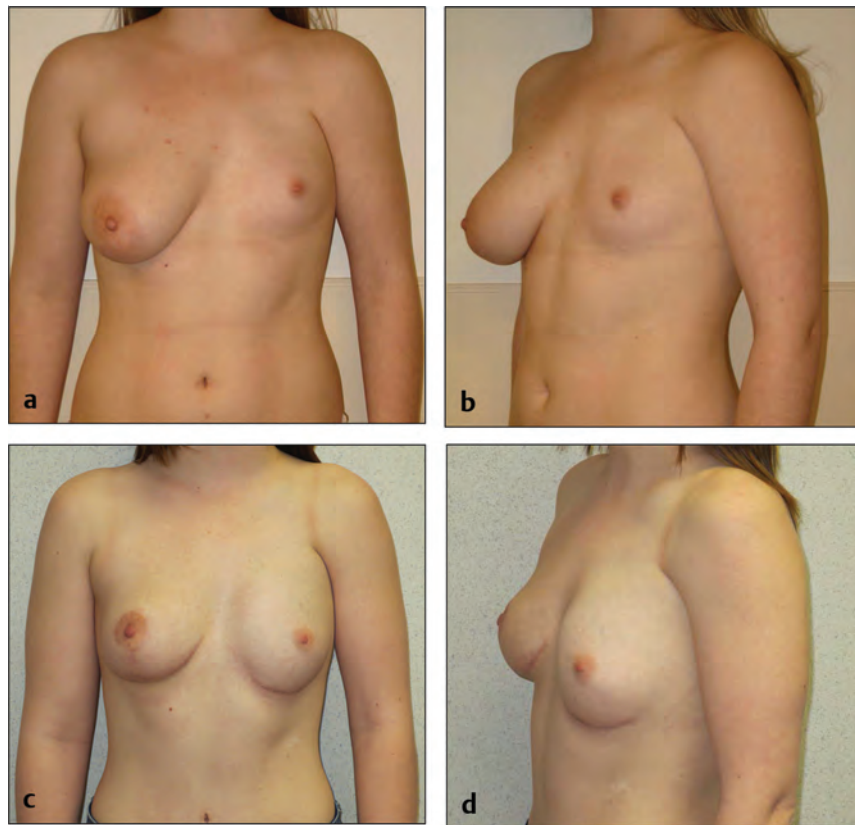


Fig. 27.4 A 20-year-old with Poland syndrome (a, b) preoperatively and (c,d) 2 years following free transverse rectus abdominis myocutaneous (TRAM) flap reconstruction and 1 year after contralateral mastopexy. Prior to free-tissue transfer, tissue expansion was used to develop an adequate skin envelope for the reconstructed breast.

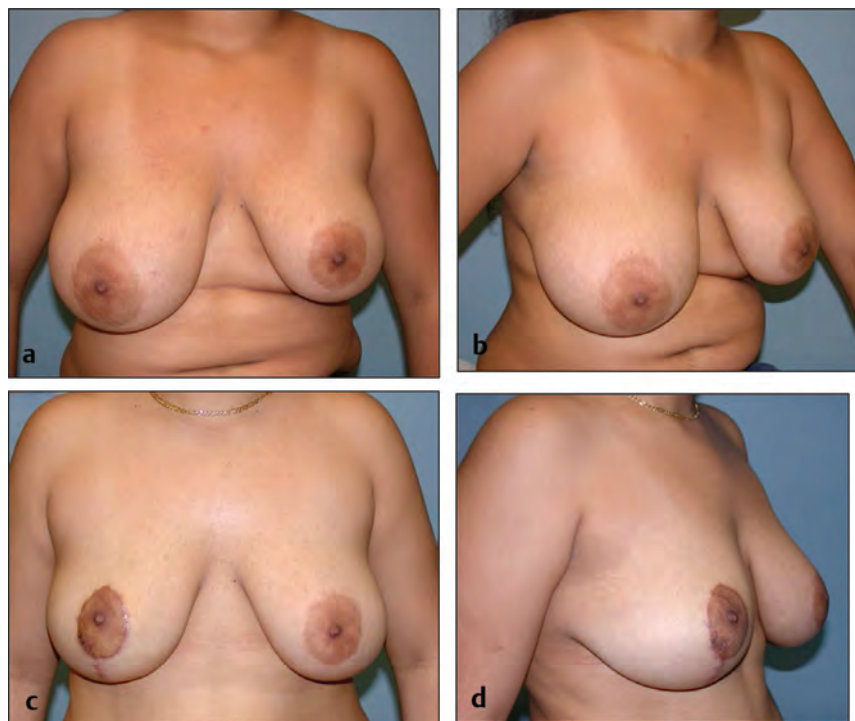


Fig. 27.5 An 18-year-old with moderate breast asymmetry (a,b) preoperatively and (c,d) 1 year following unilateral, right side vertical scar reduction mammoplasty (380 g).



Fig. 27.6 A 19-year-old with severe breast asymmetry (a,b) preoperatively and (c,d) 8 years following right Wise pattern reduction mammoplasty (1,500 g) and 7 years following left augmentation mammoplasty with a submuscular 300 mL smooth, round gel implant.

27.5 Postoperative Management

The postoperative routine varies depending on the procedure performed. In cases of unilateral or bilateral reduction/mastopexy procedures, the surgical site is often covered with steri-strips, absorbent dressings, and a surgical support bra. At the first postoperative visit, the dressing is removed and the surgical site checked. Typically, patients require 3 to 5 days of prescription pain medication and are out of school or work for roughly 1 week. Exercise and lifting should be avoided for the first 6 weeks following surgery, after which full activity can be resumed. Similarly, patients should use a supportive sports bra and avoid a bra with an underwire for 6 weeks following reduction mammoplasty. The recovery following unilateral or bilateral augmentation procedures is typically shorter in duration. Appropriate supportive bras are used, and high-exertion or high-impact activities, especially those involving the upper extremities, are limited for at least several weeks. Given the presence of the foreign body, attention to avoiding wound-healing problems and infection is paramount. Once in place, implants should be monitored annually for potential problems such as deflation, malposition, or abnormal contour.

27.6 Complications

Postoperative complications vary by the type of surgical procedure performed. Early complications following breast augmentation may include bleeding, infection, or implant exposure. Severe cases of infection may necessitate removal of the implant. Other implant-related complications include capsular contracture, visible wrinkling or rippling, displacement, or failure requiring replacement. Secondary procedures may be required as the patient ages. Additional views will be required during mammography later in life as the implant can obscure breast tissue. In rare instances, implant-associated lymphomas have been reported and some federal recommendations have

been made for interval radiographic evaluation of the implant over time.

An estimated 40 to 50% of patients undergoing reduction mammoplasty will experience a complication. These complications are typically minor and occur during early wound healing. Infection, bleeding, wound dehiscence, and seroma or hematoma requiring reoperation and evacuation are most commonly reported. Nipple and fat necroses are exceedingly rare in the healthy adolescent population.

Patients should be followed closely during the first postoperative year to assess long-term complications. Diminished nipple and breast sensation is normal during the early healing period following any breast surgery. A subset of women, however, will have partial or complete loss of breast and/or nipple sensation that persists after the first postoperative year. Scarring and keloid formation are common following large resections.

Some degree of asymmetry is to be expected following breast surgery. Patients should be advised that although surgical intervention aims to reduce the appearance of asymmetry, some breast differences may persist following surgery. Differences in breast size and position are common following implant placement. Additionally, discrepancies in NAC size and position may be appreciated after reduction mammoplasty and mastopexy. Revisional procedures to correct postoperative asymmetry should be addressed after the first year to allow swelling to resolve and the breast and/or implant to settle.

Adolescents undergoing surgical intervention may require future reoperation due to continued breast growth. In some instances, the augmented breast may outgrow the contralateral breast, or the hyperplastic breast may continue to grow after reduction. Although this is certainly possible and surgical intervention should be delayed until breast maturation is achieved, age should not be an absolute contraindication to treatment. For many patients, the psychosocial benefits of early surgical intervention far outweigh the risk of a repeat procedure later in life.

Patients with breast asymmetry may also be at an increased risk for future lactation difficulty. Breast hypoplasia by definition may curtail milk volume due to breast gland insufficiency. Surgical intervention may also diminish nipple sensitivity and impede future latching and lactation. Although reduction mammoplasty largely spares neurovascular structures of the ductal breast tissue, patients may need to supplement their milk supply. Furthermore, breast implant placement may hinder a woman's ability to lactate. Patients undergoing surgical intervention should be counseled with respect to future lactation.

27.7 Conclusion

Most women will exhibit breast asymmetry during their lifetime. Breast asymmetry is not a formal diagnosis, but rather a broad description encompassing both normal and abnormal differences in breast size, shape, or position. Size and shape differences are particularly prevalent during early breast maturation. Most adolescent cases of breast asymmetry are due to variable rates of breast gland proliferation during early puberty and will improve or resolve completely without intervention. Less commonly, asymmetry may be secondary to hyperplastic and hypoplastic breast conditions of developmental or congenital origin.

Asymmetry of any severity can negatively affect a patient's quality of life, self-esteem, and social functioning. Early intervention, however, may alleviate and prevent the exacerbation of psychosocial symptoms into adulthood. Young patients who are concerned by the appearance of their breasts should be encouraged to explore prostheses or bra inserts to improve the appearance of their chest within clothing. Surgery may be warranted in cases of abnormal asymmetry that persists into late adolescence or early adulthood. A combination of breast augmentation, reduction mammoplasty, mastopexy, and tissue expansion can be employed to achieve symmetry with the contralateral breast and meet the individual goals of the patient.

27.8 Key Points

- Breast asymmetry is a common finding in adolescent and adult women.
- Breast asymmetry describes normal and abnormal differences in breast size, shape, and position.
- The majority of adolescent cases are idiopathic and self-limiting.
- Less commonly, asymmetry may be secondary to hyperplastic or hypoplastic breast conditions, benign or neoplastic masses, trauma to the breast bud or gland, or prior breast surgery.
- Surgical treatment may be warranted for distressing, abnormal, or persistent asymmetry, and can comprise a combination of breast augmentation, mastopexy, and reduction mammoplasty.

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28 Anterior Trunk Disorders

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Summary

Disorders of the anterior trunk include congenital defects of the abdominal or chest wall. Gastroschisis and omphalocele are the two most common congenital abdominal wall defects. The absence of a covering membrane around the herniated abdominal contents differentiates gastroschisis from omphalocele, and also may affect timing of surgical management. Bladder exstrophy is a rare malformation of the infra-umbilical abdominal wall that is easily recognizable at birth, but is rarely diagnosed prenatally. Prune belly syndrome is characterized by hypoplastic or absent anterior abdominal wall musculature leading to a wrinkled appearance of the abdomen. Congenital chest wall diagnoses include pectus excavatum and pectus carinatum. Pectus excavatum is characterized by a concave appearance to the anterior chest wall, while pectus carinatum presents as a protrusion of the sternum and/or ribs with a depression along the sides of the chest.

The goal of surgery for patients with gastroschisis and omphalocele is to safely return the eviscerated bowel to the peritoneal cavity and establish abdominal wall integrity. Surgical closure for gastroschisis and omphalocele can be achieved via primary reduction, or delayed, staged closure. Treatment for bladder exstrophy usually occurs in a staged manner to close the bladder and abdominal wall. Plastic surgeons may be involved in late corrections of abdominal scarring and, incisional hernias or bulges. Although the majority of umbilical hernias will close spontaneously, large defects or those that persist to school age may require surgical repair. A variety of non-operative, minimally invasive and invasive modalities exist to treat both pectus carinatum and excavatum.

Keywords: anterior trunk disorders, gastroschisis, omphalocele, bladder exstrophy, Prune belly syndrome, pectus excavatum, pectus carinatum, congenital

28.1 Introduction

Disorders of the anterior trunk constitute a wide array of congenital defects of the abdominal or chest wall. These disorders range in severity and frequency and are typically managed by pediatric surgeons. Plastic surgeons who treat children and young adults can apply a host of reconstructive and aesthetic techniques in the early as well as late management of these conditions. As such, it is worthwhile for the plastic surgeon to familiarize themselves with some of the more common anterior trunk disorders.

Minor congenital abdominal wall defects are common. In particular, umbilical or inguinal hernias are frequently encountered. Gastroschisis and omphalocele are the most common serious congenital abdominal wall defects, and are frequently diagnosed antenatally. Gastroschisis is characterized by free herniation of the abdominal viscera. There is no membrane covering the herniated abdominal contents, which differentiates gastroschisis from omphalocele. This condition has doubled in incidence over the past decade for unclear reasons, with

approximately 2,000 new cases per year in the United States. Omphalocele is a related condition also characterized by evisceration of intra-abdominal organs. However, these viscera are contained in a sac that protects the viscera, minimizes neonatal fluid losses, and affords substantial time to repair unlike gastroschisis (► Fig. 28.1). Unlike the periumbilical defects above, bladder exstrophy is a rare malformation of the infraumbilical abdominal wall. Incomplete closure of the bladder, protrusion of the posterior bladder wall through the lower abdominal wall, epispadias, as well as alterations in the pelvic bones and muscles necessitate early acute multidisciplinary care. Late defects of the abdominal wall such as bulges, hernias, and depressed scars are common. In addition, complex genitourinary defects may also require multiple complex reconstructive procedures. Another very rare condition that affects both the abdominal wall and the urinary tract is prune belly syndrome. In this condition, the anterior abdominal wall musculature is hypoplastic or absent, leading to a wrinkled appearance of the abdomen (► Fig. 28.2). In addition to the marked skin redundancy and distortion of the trunk, severe urological abnormalities and cryptorchidism frequently occur.

The range of a congenital chest wall diagnoses is smaller, but some diagnoses are common as well. Pectus excavatum, for example, may occur in 1/150 to 1/1,000 live births and is familial in 40% of cases. It is characterized by a concave appearance to the anterior chest wall and is the most common chest wall abnormality. The depression may be mild or quite severe, impacting cardiac and pulmonary function (► Fig. 28.3). In addition, it can be present at birth or manifest during adolescence. In contrast, pectus carinatum presents as a protrusion of the sternum and/or ribs with a depression along the sides of the chest (► Fig. 28.4). Though not as common as pectus excavatum (occurring only 1/10th as often), it accounts for roughly 5 to 7% of chest wall anomalies overall. The condition is usually isolated and nonsyndromic though pectus carinatum can be



Fig. 28.1 A 1-month-old male with large omphalocele. Note the well-epithelialized sac containing the viscera.



Fig. 28.2 A 13-year-old male with prune belly syndrome. Note urostomy in the right lower quadrant and scarring from prior urological procedures.

seen in patients with Marfan, Ehlers–Danlos, Noonan, and Turner syndromes as well as others.

28.2 Diagnosis

Most diagnoses of the anterior trunk can be made following history and physical examination. Radiological evaluation may also be extremely useful early or even later in life. Severe conditions can also be recognized by prenatal ultrasound and appropriate plans for postdelivery management made in advance. In many of these conditions, concurrent severe comorbidities may exist and must be sought out.

The prenatal diagnosis of gastroschisis is possible from the end of the first trimester, after the physiological closure of the abdominal wall around 10 weeks of gestation. The ultrasound detects loops of bowel outside of the abdominal cavity, herniated through a small paraumbilical wall defect and floating in the amniotic fluid without any covering membrane. The absence of this covering allows one to differentiate gastroschisis from omphalocele. Present techniques can predict complicated gastroschisis and outcomes associated with degree of intra-abdominal dilatation. A raised maternal alpha fetoprotein is also indicative of gastroschisis. Gastroschisis is not associated with chromosomal anomalies and generally occurs as an isolated anomaly. An omphalocele can also be diagnosed by ultrasound during the late first trimester, but is more commonly



Fig. 28.3 A 16-year-old female with mild pectus excavatum.



Fig. 28.4 A 14-year-old male with mild pectus carinatum.

confirmed during the 18-week ultrasound. In some cases, an elevated maternal alpha fetoprotein level may also be identified, though less commonly than in gastroschisis. Associated anomalies are common and include cardiac, renal, skeletal, and neural tube abnormalities. Omphaloceles are often associated with chromosomal anomalies (such as Trisomy 13, 18, and 21) as well as other syndromes such as Beckwith–Wiedemann.

Umbilical hernias occur when the umbilical ring is not fully closed and can be diagnosed upon physical examination. Rarely is any imaging required. A protruding soft-tissue mass and a palpable defect in the fascia is diagnostic. Similarly, pediatric inguinal hernias are diagnosed clinically with a visible soft-tissue bulge in the inguinal region. In boys, ipsilateral scrotal swelling may be secondary to a hernia or a hydrocele. Ultrasoundography may be used to differentiate between the two.

Bladder exstrophy is easily recognizable at birth, but is rarely diagnosed prenatally. The prenatal diagnosis consists of a non-visible fetal bladder and low insertion of the umbilical cord. Prune belly syndrome can be diagnosed through a prenatal ultrasound, although diagnosis is usually made shortly after birth. It occurs most often in male infants. The ultrasound diagnosis in early gestation requires the identification of a megacyst (distended bladder) and oligohydramnios. Undescended testes and urinary tract anomalies are often noted postnatally on physical examination and imaging.

The pectus excavatum and carinatum deformities are easily visualized on physical examination. In some, the deformity may be present early in life, whereas in others it will manifest during adolescent growth. Symptoms may include exercise intolerance, decreased endurance, pain, body image issues, and dyspnea. In symptomatic individuals, computed tomography (CT) scans may be warranted to determine the severity of the pectus defect and potentially assist in reconstructive planning. In cases of pectus excavatum, cardiac and pulmonary compression may be documented in conjunction with functional tests such as an echocardiogram or pulmonary function testing. Patients with pectus carinatum manifest a sternal protrusion with a depression along the sides of the chest. Most are male and present during adolescence. Associated symptoms including chest wall pain, frequent injuries, body image issues, and scoliosis should be sought. Displacement of the thoracic musculature can interfere with breathing, and 10% of patients experience exercise intolerance. In symptomatic patients, a chest CT scan in conjunction with pulmonary function testing can document severity.

28.3 Nonoperative Treatment

Many congenital defects of the anterior trunk may benefit by nonoperative treatment either alone or in combination with subsequent operative treatment. Although both gastroschisis and omphalocele ultimately require surgical treatment, nonsurgical prenatal and neonatal care is equally essential in the management of these patients. In particular, body temperature and intravascular volume need to be maintained. In addition to intensive neonatal care, appropriate positioning and dressing applications are essential to avoid kinking of the mesentery or viscera to minimize the risk of ischemic injury.

Unless incarcerated or producing systemic symptoms, umbilical and inguinal hernias do not require urgent surgical intervention. In the case of umbilical hernias, if the defect is small (<1 or 2 cm), 90% close within 3 years and some sources state 85% of all umbilical hernias, regardless of size, will close without surgical treatment. In contrast, inguinal hernias do not spontaneously heal and should be repaired operatively.

Patients with bladder exstrophy and those with prune belly syndrome will present with both medical and surgical needs. Just as with omphalocele and gastroschisis, neonatal as well as

intensive ongoing medical care is essential for these patients early in life.

The majority of patients with pectus deformities are minimally symptomatic or asymptomatic. In many cases, support and reassurance may be all that is required. In the case of pectus carinatum, there have been reports of bracing being used to help diminish the prominence of the sternum.

28.4 Operative Treatment

28.4.1 Gastroschisis

The goal of surgery is to safely return the eviscerated bowel to the peritoneal cavity and establish integrity to the abdominal wall. The absence of any protective membrane over the bowel as in omphalocele mandates urgent coverage and vigilant fluid and temperature maintenance for the neonate. The degree of visceroperitoneal disproportion will determine whether the reduction can be performed primarily or in a staged manner. Unless the defect is very small, a Silastic “silo” or cone is sewn to the periphery of the fascial defect and sequential daily compression is used to reduce the viscera progressively over time. As the viscera are reduced, ventilation, intra-abdominal pressure, and venous return will all be affected. It is vital to monitor these parameters as staged reduction occurs to avoid major morbidity and mortality. If the defect is so large or the patient's physiology so marginal, direct fascial repair may not be possible even with a staged approach over days. The pediatric plastic surgeon may be of assistance utilizing abdominal reconstructive modalities such as biologic or prosthetic mesh, flap coverage, or component separation to achieve temporary closure. Once the child has aged, secondary reconstruction by the plastic surgeon can often improve both the appearance and the function of the abdominal wall.

28.4.2 Omphalocele

The treatment goals and risks for patients with omphalocele are similar to those with gastroschisis. However, there are some very important differences in timing that need to be emphasized. The presence of a membrane over the herniated viscera is protective and thus allows for a considerably longer timeline for reduction. In fact, successful treatment of large omphaloceles has been extended over years. Thus, it is paramount to preserve the integrity of the omphalocele sac, which will epithelialize over a couple of weeks. If the defect is small or the degree of visceroperitoneal disproportion is minimal, early closure can certainly be achieved. However, the presence of medical comorbidities, especially cardiac anomalies, must be accounted for along with the potential sequelae discussed earlier. The availability of additional time for closure allows the reconstructive surgeon the option for tissue expansion in addition to the other treatment modalities discussed earlier (► Fig. 28.5). Subcutaneous, submuscular, as well as intraperitoneal tissue expansion have all been reported in successful omphalocele closure.

28.4.3 Hernia

Although the majority of umbilical hernias will close spontaneously, large defects or those that persist to school age may



Fig. 28.5 An 8-year-old female with large, persistent umbilical hernia.

require surgical repair (► Fig. 28.6). Most repairs will be managed by the pediatric surgeon. However, those with significant skin excess or umbilical distortion may be referred for plastic surgical assistance. Inguinal hernias invariably required surgical repair. Fortunately, recurrence rates are low, and the procedure is typically well tolerated. It is unusual for plastic surgical techniques to be required for these defects.

28.4.4 Bladder exstrophy

Cloacal or bladder exstrophy requires considerable multispecialty surgical care and medical management. Treatment usually occurs in a staged manner to close the bladder and abdominal wall as well as repair the bladder neck, and ultimately the epispadias. Although plastic surgeons are rarely involved early in the patient's life, late corrections of abdominal scarring, incisional hernias, or bulges can substantially improve these patients' quality of life later on. Furthermore, genitourinary complications such as urinary incontinence from incomplete bladder neck closure or genital deformities can also be addressed with plastic surgical techniques to assist the pediatric urologist.

28.4.5 Prune Belly Syndrome

Patients with prune belly syndrome typically demonstrate some degree of abdominal distension and skin wrinkling secondary to the aberrant and underdeveloped abdominal musculature. Although renal and genitourinary issues predominate early in life, adolescents and young adult patients may benefit from abdominal wall reconstruction to advance the well-innervated and well-vascularized peripheral muscle layers centrally along with excision or plication of the deficient central muscle and fascial tissues. This can be approached as for an abdominoplasty, and excision of redundant skin can also be performed at the same time. A variety of techniques have been reported in the plastic surgical literature.

28.4.6 Pectus Excavatum and Pectus Carinatum

Surgical treatment of pectus deformities can be used to address functional as well as aesthetic concerns. Historically, pediatric surgeons have used two surgical techniques to address pectus excavatum deformities, although newer alternative modalities have certainly been described. The Ravitch procedure introduced in the mid-20th century is an open procedure in which the abnormal rib cartilage is resected while preserving the perichondrium. The sternum is repositioned and held in place with a plate until the cartilage has reformed. The Nuss procedure is a minimally invasive alternative in which a metallic bar is introduced intrathoracically via two small incisions. The bar is then rotated into position adopting a convex posture to reposition the costal cartilages and sternum. The bar is typically removed 2 to 5 years later. Patients without symptoms referable to the deformity or those who simply wish to improve the appearance of their chest may be referred to a plastic surgeon. A variety of fill options including structural fat grafting and custom-made implants have been described.

Pectus carinatum is usually corrected during adolescence or late childhood. Although bracing options exist, a great deal of compliance and dedication is required to complete therapy. For those patients who fail bracing, similar surgical options (e.g., modified Nuss or Ravitch procedure) to pectus excavatum can be employed. Camouflage options such as breast or chest augmentation also exist for carinatum patients as well. In general, outcomes for surgical treatment of both pectus deformities range from good to excellent, with postoperative improvement noted in exercise tolerance, quality of life, and body image perception.

28.5 Complications

Surgical complications following treatment of anterior trunk deformities range from minor to major, and reflect the type of surgical procedure performed. Major complications following an excessively tight or rapid gastroschisis or omphalocele closure include abdominal compartment syndrome, necrotizing enterocolitis, sepsis, renal failure, cardiopulmonary failure, and enterocutaneous fistula. Late complications include hernias, bulges, and hypertrophic or depressed scars. Complications following umbilical or inguinal hernia repair are infrequent and generally minor. Although spermatic cord or bowel injuries can

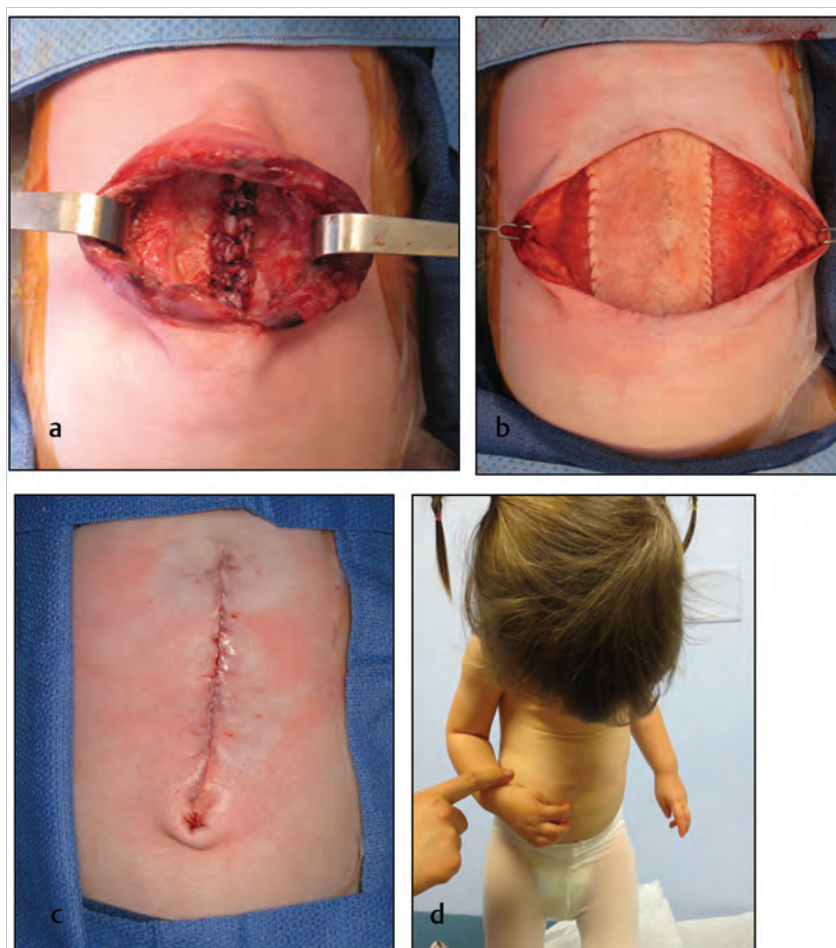


Fig. 28.6 (a) A 9-month-old female with large omphalocele treated with component separation to allow for fascial closure. (b) Onlay acellular dermal matrix over suture line. (c) Closure and umbilicoplasty. (d) One-year follow-up.

occur after inguinal hernia repairs, these major complications are very rare. Either umbilical or inguinal hernias can recur and may require additional surgery. Exstrophy patients may suffer similar complications to patients with other abdominal wall defects following early surgical procedures. Long-term complications such as recurrent urinary tract infections, incontinence, sexual dysfunction, and pain are also common. The surgical complications following surgery for prune belly syndrome are frequent due to the high rate of renal and pulmonary dysfunction and are similar to risks for abdominoplasty, such as seroma, hematoma, dehiscence, and tissue loss.

Surgical complications following pectus repair also range from minor to major depending on procedure type. Potential complications in the management of pectus excavatum include recurrence, floating sternum, breast hypoplasia, postoperative pain (more common with Ravitch procedure), and implant migration/extrusion with custom implants. Complications in the treatment of pectus carinatum are similar to that of pectus excavatum and include treatment failure in the case of bracing or recurrence if a conservative resection is performed or the support bar is removed prematurely.

28.6 Conclusion

Defects of the anterior trunk are common and encompass a wide range of diagnoses. Although major defects such as

gastroschisis are rare, the incidence may be increasing. Most major defects are now diagnosed antenatally and allow for adequate time to prepare the parents and medical and surgical teams in advance of delivery. Minor defects such as hernias or mild pectus deformities are far more common and diagnosed postnatally or later in childhood during physical examination. For some diagnoses, nonsurgical or minimally invasive options exist. In others, nonsurgical techniques can facilitate subsequent operative procedures. The reconstructive background of the plastic surgeon allows for a unique role in both the acute and late management of these conditions. For example, familiarity with fascial substitutes, component separation, and tissue expansion can facilitate closure of large abdominal wall defects. Similarly, application of aesthetic principles to patients who have undergone exstrophy correction earlier in life or who have prune belly syndrome can similarly improve self-esteem and quality of life. Although involvement varies from center to center, there are probably many patients with anterior trunk disorders that could benefit from plastic surgical evaluation and treatment who are not afforded the opportunity to do so.

28.7 Key Points

- Gastroschisis and omphalocele are the most frequent major congenital abdominal wall defects. They differ in terms of urgency to achieve closure and potential comorbidities.

- For larger abdominal wall defects, biological or prosthetic mesh, component separation, and tissue expansion may be required.
- Plastic surgeons can offer patients with prune belly syndrome and bladder exstrophy improved quality of life with secondary reconstructive procedures later in adolescence or young adulthood.
- Pectus excavatum and pectus carinatum are two common chest wall deformities that may alter cardiopulmonary physiology or may simply be an aesthetic concern. A variety of non-operative, minimally invasive, and invasive modalities exist to treat both anomalies.

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29 Posterior Trunk Disorders

Arin K. Greene

Summary

Posterior trunk disorders can be congenital (e.g. myelomeningocele) or acquired (e.g. pressure ulcers), and frequently require the expertise of plastic surgeons. Soft-tissue reconstruction is often required for wounds over the spine. Typically these defects are closed with local tissue; grafts and free-tissue transfer rarely are necessary.

Diagnosis of posterior trunk disorders is principally made by history and physical examination. Management is influenced by the depth of the wound, exposure of dura, infection, wound size and location, presence of hardware, duration of the wound, sensation and ambulatory status of the patient. The underlying cause of the wound must be addressed before considering operative interventions. For instance, pressure ulcers typically heal without operative intervention once the excessive pressure has been eliminated.

Myelomeningocele repair usually involves neurosurgical intervention in the first two days of life. Once the dura is closed, a plastic surgeon may be required to provide soft-tissue coverage for large defects. The reconstruction occurs in two planes: flaps over the dural repair, and skin closure. Regional muscles, perforator flaps, skin grafts, and skin flaps are all potential options.

The first line of management of pressure ulcers in the pediatric population involves periodic weight shifting, and specialized wheel-chair cushions and beds for non-ambulatory patients. It is important to investigate the cause of the wound and to eliminate the etiology. Whilst conservative therapy is the mainstay of treatment, operative closure is sometimes required. Other posterior trunk disorders, such as wounds over the spine and traumatic soft-tissue defects, are also managed by plastic surgeons.

Keywords: posterior trunk disorder, myelomeningocele, pressure ulcer, soft-tissue, reconstruction, spine

29.1 Introduction

Posterior trunk disorders can be congenital or acquired. The most common malformation plastic surgeons are asked to manage is myelomeningocele. The most frequent acquired defect is a pressure ulcer. Plastic surgeons often are asked to reconstruct soft tissue for wounds over the spine. Generally, posterior trunk defects are able to be closed using local tissue; grafts and free-tissue transfer rarely are necessary.

29.2 Diagnosis

Diagnosis of posterior trunk disorders is made by history and physical examination. Imaging and histopathology are rarely indicated. Variables that will dictate treatment include the following: (1) depth of wound, (2) exposure of dura, (3) infection, (4) wound size and location, (5) presence of hardware, (6) duration of the wound, and (7) sensation and ambulatory status of the patient. If a wound has been present for at least 6 weeks, then it is chronic and likely has not healed because of infection, bacterial colonization of hardware, or pressure. The underlying

cause of the wound must be addressed before considering operative interventions.

29.3 Nonoperative Treatment

The surgeon managing a posterior trunk disorder should begin considering reconstructive options as low on the reconstructive ladder as possible. Pressure ulcers typically heal without operative intervention once the excessive pressure has been eliminated. Superficial wounds can be allowed to heal secondarily. Before considering operative intervention, the benefits of the procedure must outweigh its risks.

29.4 Operative Treatment

29.4.1 Myelomeningocele

Myelomeningocele is the most common neural tube defect affecting approximately 1/3,000 children. The spinal cord is exposed due to a cleft in the vertebral column and overlying soft tissue. The repair is usually performed on the first or second day of life to prevent cord desiccation and meningitis. After the neurosurgeon detethers the spinal cord, infolds the placode, and closes the dura, he/she may ask for a plastic surgeon to provide soft-tissue coverage if the defect is large. Several methods have been described to reconstruct tissue over the dural repair: regional muscles (latissimus dorsi, gluteus maximus), perforator flaps, skin grafts, and skin flaps (bilobed, bipedicle, rhomboid, rotation, V-Y advancement).

I believe the best method to manage a myelomeningocele is to reconstruct the area in two planes: (1) flaps over the dural repair and (2) skin closure (Box 29.1). First, I use turnover paraspinous fascial flaps to cover the dural repair (► Fig. 29.1 and ► Fig. 29.2). The flaps are elevated laterally to medially, turned over, and sutured in the midline with interrupted 4–0 vicryl. Hydrodissection with local anesthetic lifts the fascia off the underlying muscle facilitating the dissection. The paraspinous fascia provides a strong, tension-free, vascularized layer over the dural repair. The thick tissue reinforces the dural incision line to reduce the risk of cerebrospinal fluid leak. The fascial flaps separate the dural and skin closure to reduce the risk of cerebrospinal fluid contact with the integument and meningitis.

Box 29.1 Principles of Extradural Myelomeningocele Reconstruction

1. Paraspinous turnover fascial flaps
 - a) Cover the dura and decrease cerebrospinal fluid leak
 - b) Separate dural repair from skin closure
 - c) Avoid gluteal/latissimus flaps, which can defunctionalize a neurologically compromised patient
2. Linear midline skin closure
 - a) Reduces complications
 - b) Facilitates reoperations for tethered cord and scoliosis

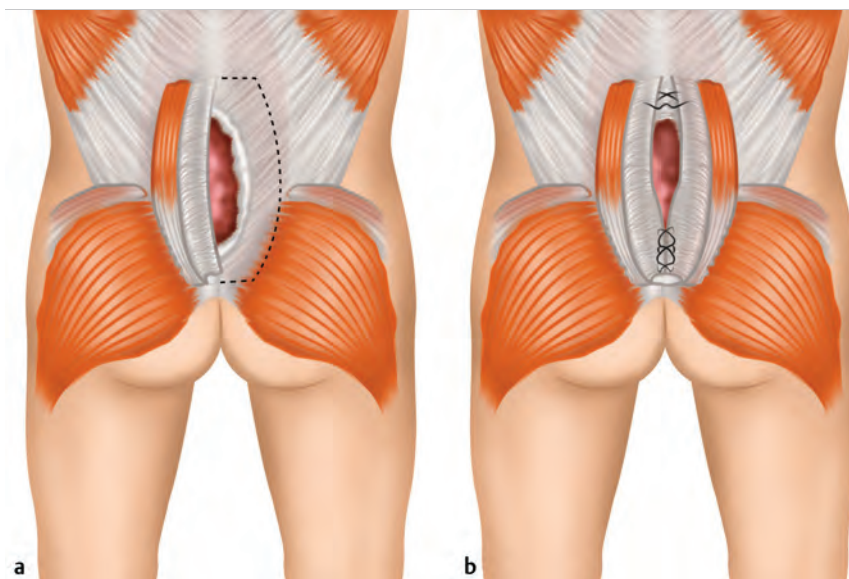


Fig. 29.1 Extradural myelomeningocele reconstruction. (a) Fascia is incised laterally and elevated medially over the underlying muscles. (b) The turnover fascial flaps are sutured together in the midline. (Adapted from Patel KB, Taghinia AH, Proctor MR, Warf BC, Greene AK. Extradural myelomeningocele reconstruction using local turnover fascial flaps and midline linear skin closure. *J Plast Reconstr Aesthet Surg* 2012;65:1569–1572.)

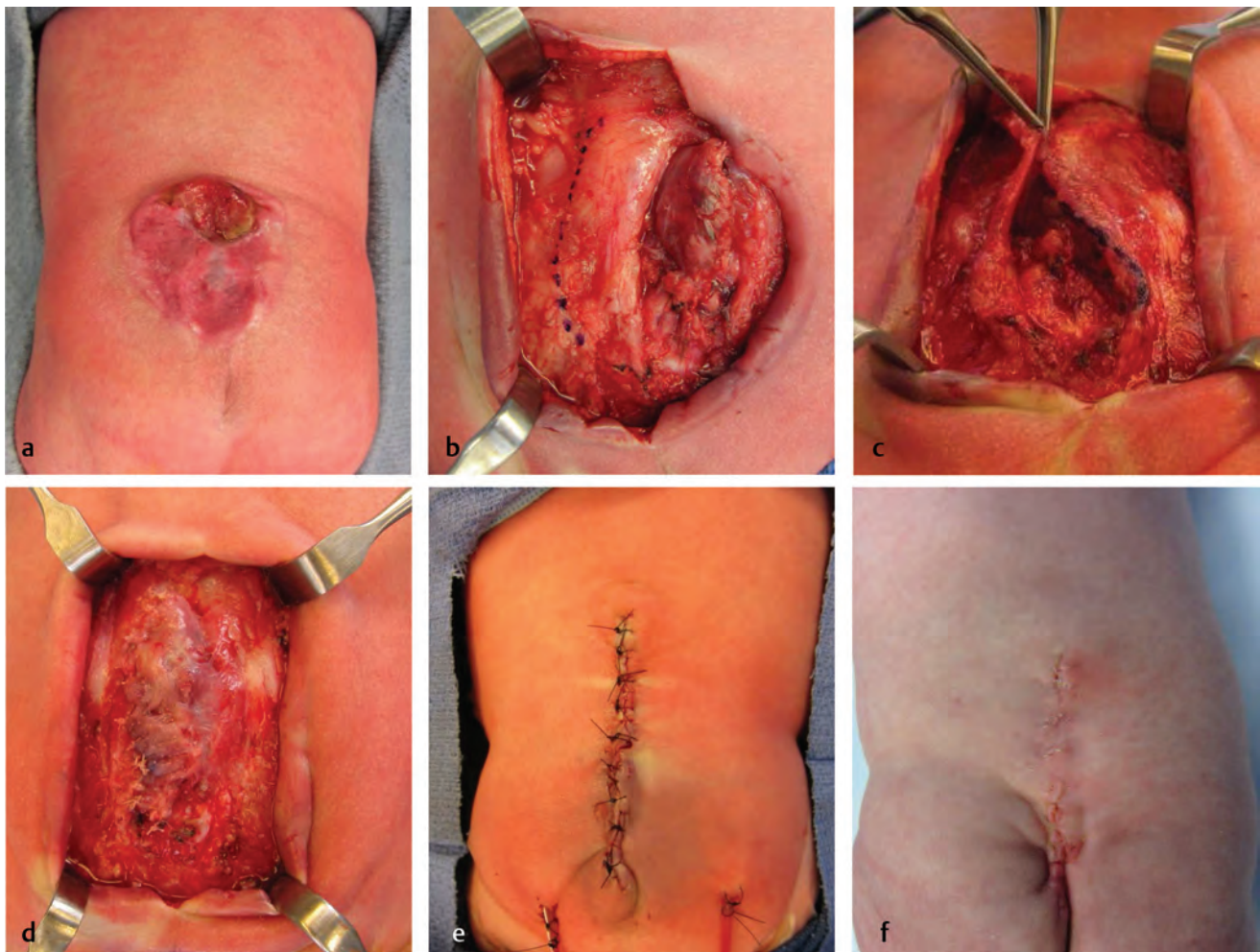


Fig. 29.2 A 1-day-old male with a $6 \times 5 \text{ cm}^2$ myelomeningocele. (a) Preoperative appearance. (b) Lateral incision marked to incise the paraspinal muscle fascia. (c) Bilateral turnover fascial flaps raised. (d) Flaps sutured together over dura. (e) Skin defect closed linearly with drains after widely undermining skin flaps. (f) Following suture removal 6 weeks postoperatively.

This technique does not include muscle dissection and thus will not worsen the functional deficit in a neurologically compromised patient. The patient's ambulatory status is related to the level of spinal involvement. Ninety-five percent of children with L5 or sacral myelomeningocele are able to ambulate moderate distances (community ambulators). Patients with defects at L3 or above are only able to ambulate short distances (nonfunctional ambulators). Sixty percent of patients with myelomeningoceles will be community ambulators, 15% nonfunctional ambulators, and 25% are nonambulatory. Paraspinous turnover fascial flaps obviate the need to use major muscle flaps (e.g., gluteal, latissimus), which are more complicated to dissect and have a greater risk of complications. Harvest of gluteal and/or latissimus muscles also can further defunctionalize an infant with a myelomeningocele. Use of gluteal muscle flaps in a potential community ambulator may cause the child to become a nonfunctional ambulator or nonambulatory. Harvest of latissimus muscle will handicap a child's ability to walk with crutches and/or transfer from a wheelchair.

After providing deep soft-tissue coverage over the dural repair with paraspinous fascia, the overlying skin is approximated. To approximate the integument, skin flaps are widely undermined above the muscle fascia, advanced medially, and sutured vertically in the midline over two drains with interrupted 4-0 vicryl and 4-0 nylon suture. If large defects are closed with tension, prone positioning is prescribed for a minimum of 2 weeks postoperatively and the nylon sutures are maintained for at least 4 weeks.

Skin closure in the midline is important for myelomeningocele repair because patients often require additional operations involving the posterior trunk. Secondary procedures for cord tethering or spinal fusion for scoliosis occur in one-half of patients. I avoid the use of bilobed, bipedicle, rhomboid, rotation, or V-Y advancement flaps to minimize posterior trunk scarring and complications (► Fig. 29.3). The vertical scar facilitates exposure for secondary spinal procedures and minimizes the risk of skin necrosis from multiple posterior trunk incisions. Wide skin undermining will achieve a linear, midline closure without breakdown for even the largest defects. If the incision

line separates, it will heal secondarily without consequence; the risk of meningitis is small because fascial flaps separate the integument from the dura.

29.4.2 Pressure Ulcer

Sacral and ischial pressure ulcers occur in children who are insensate (usually from myelomeningocele) or who have significant developmental delay and are nonambulatory. Pressure ulcers in the pediatric population are managed similarly to adults. Patients/providers are instructed to shift their weight every 2 hours and are prescribed wheelchair cushions and beds that reduce the risk of pressure ulcers.

Initial management of a pressure ulcer is to investigate what caused the wound and to eliminate the etiology. Often, a change in a cushion, new wheelchair, or a recent hospitalization cause the skin breakdown. Full-thickness wounds are managed conservatively with dressing changes; necrotic tissue is sharply debrided. A wristwatch set to alarm every 2 hours will remind the patient/provider to shift the patient's weight. Almost all wounds will heal with local wound care and elimination of pressure.

Exposed bone (stage 4) does not equate with osteomyelitis, which is very rare. Patients with large wounds have increased inflammatory markers. Magnetic resonance imaging (MRI) is not a useful modality for diagnosis of osteomyelitis and will show inflammatory changes because of the overlying wound. Definitive diagnosis of osteomyelitis requires bone biopsy. Wounds will contract and close over exposed bone. A vacuum-assisted wound closure device may speed the healing of pressure ulcers and is an easier dressing regimen compared to gauze. Before placing a vacuum-assisted device, the area must be clean of necrotic tissue.

If a wound does not improve after several weeks, then the underlying pressure has not been adequately addressed. Closing the wound operatively will fail if the patient/provider has not eliminated pressure. Often, patients want a "quick-fix" and operative closure of the wound. However, they must be educated that if they do not eliminate pressure the defect will recur

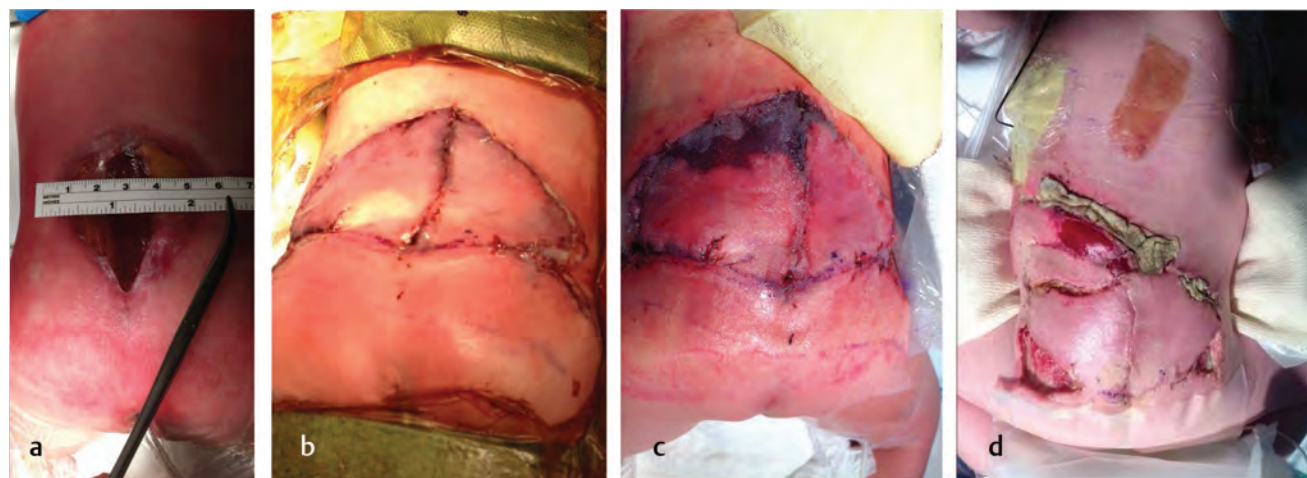


Fig. 29.3 Myelomeningocele repaired at an outside hospital with V-Y flaps. (a) Preoperative appearance. (b) Following closure of the soft-tissue defect. (c) Skin necrosis of flaps. (d) Full-thickness skin loss.



Fig. 29.4 Chronic wound following incision and drainage of an infected incision following an orthopaedic spine procedure. **(a)** Preoperative appearance. **(b)** Because the wound was superficial, it was closed with delayed primary closure after widely undermining the skin.

following any procedure. Large wounds that have taken a long time to heal can develop an epithelialized sinus tract. This is an indication to provide operative closure. The tract is painted with methylene blue and then excised. Local muscle is approximated to eliminate dead space and skin is advanced and closed linearly. Complicated flaps are not required.

29.4.3 Coverage of Spinal Wounds

Wounds over the midline spine usually result from incision line dehiscence following a neurosurgical or orthopaedic procedure. Principles of repair are the same as for myelomeningocele. If the dura is exposed, it is best to turnover paraspinous fascial flaps prior to closure of the integument; this usually occurs if the wound follows cord detethering. If the wound only involves the skin, then the integument can be approximated linearly (► Fig. 29.4). Occasionally titanium hardware is exposed from a spinal fusion for scoliosis. If this occurs, then gluteal and/or latissimus muscle flaps may be required to provide additional soft tissue to cover the hardware to prevent exposure (► Fig. 29.5). In some cases, it is best to temporarily remove spinal hardware, obtain definitive wound closure, and then reapply the hardware under stable soft tissue.

Skin defects following wound dehiscence usually can be closed linearly with delayed primary closure, especially because there is no skin loss. The posterior trunk is a favorable location to allow tissues to heal secondarily. Delayed primary closure can be performed easily by widely undermining skin flaps and suturing them in the midline. Similar to myelomeningocele, repair of posterior trunk wounds should be done with a single vertical line. Rotation/transposition of skin flaps or skin grafts should be avoided because there is a higher risk of wound healing problems and difficulty accessing the spine if additional procedures are necessary in the future.

The posterior trunk is at higher risk for wound dehiscence because (1) the patient is lying on the incision line and (2) there is tension on the sutures when the patient changes position. Large cutaneous sutures should be placed and not removed before 4 weeks, when the wound has 40% of its strength. Typically, the goal of the reconstructive procedure is to obtain a healed wound; suture marks are a secondary concern.

29.4.4 Trauma

The posterior trunk is a favorable location to reconstruct traumatic soft-tissue defects because (1) large areas can be allowed to heal secondarily without the risk of morbidity from contraction, (2) significant local muscle flaps are available, and (3) skin redundancy/elasticity allows wide undermining to close wounds. Generally, patients should be managed low on the reconstructive ladder; skin grafts and complicated procedures should be avoided if possible (► Fig. 29.6).

29.5 Complications

The most common complication following posterior trunk wound closure is dehiscence of the incision line. When patients are supine, pressure on the incision line can cause separation of the wound edges. In addition, bending or rising from the supine position applies tension to the sutures. Consequently, large interrupted sutures should be placed and not be removed until adequate scar strength has been achieved. If wide skin undermining has been performed causing ischemic flaps (e.g., myelomeningocele repair), then the patient is placed in the prone or lateral position for at least 2 weeks postoperatively. Wound dehiscence can also be minimized by placing drains under widely undermined skin. The drains help reduce the presence

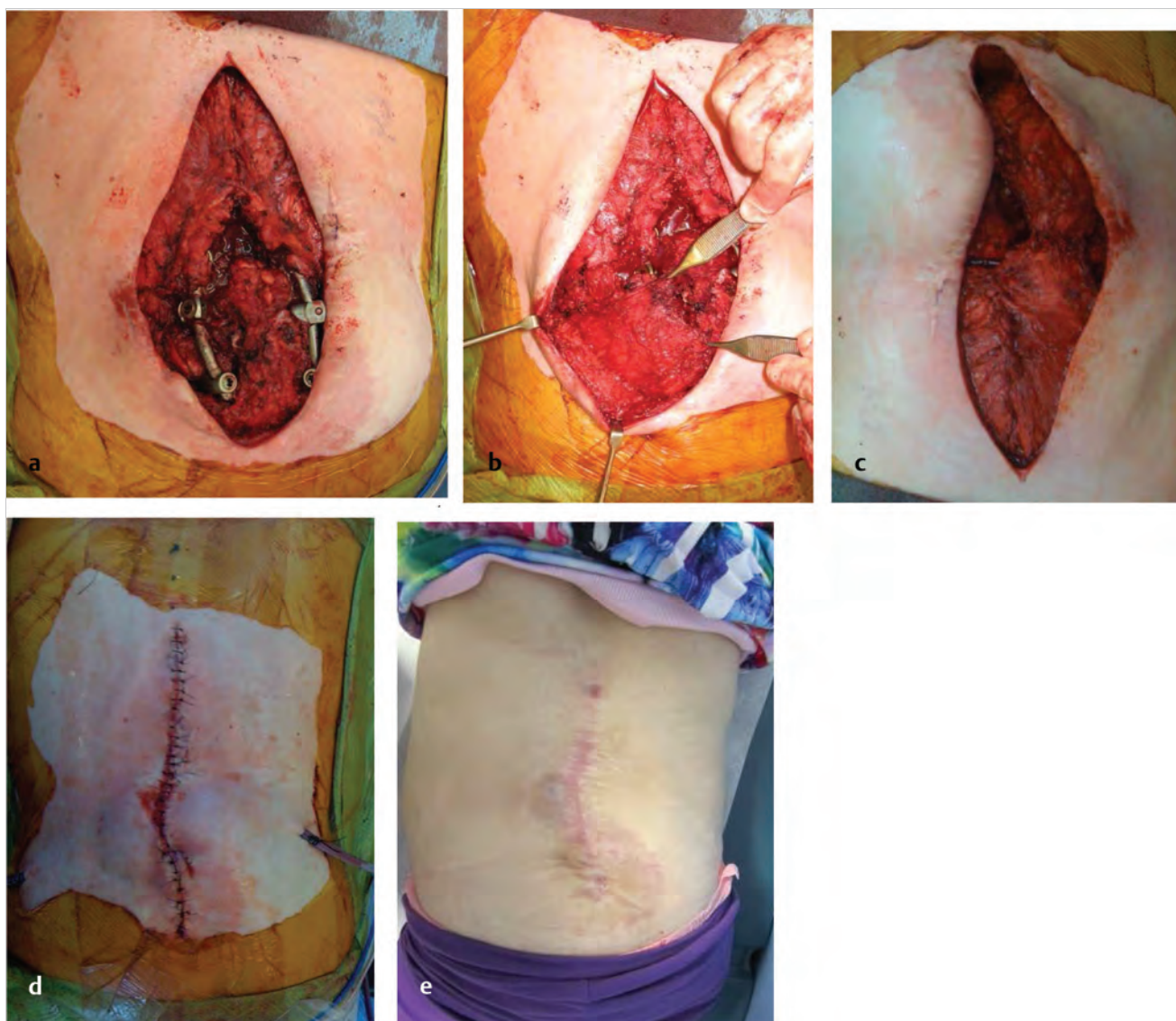


Fig. 29.5 Nonambulatory patient with myelomeningocele who developed exposed hardware following a scoliosis procedure. (a) Preoperative appearance. (b) Gluteal muscle mobilized. (c) Muscle coverage over hardware. (d) Linear midline closure after widely undermining skin flaps. (e) Three months postoperatively.

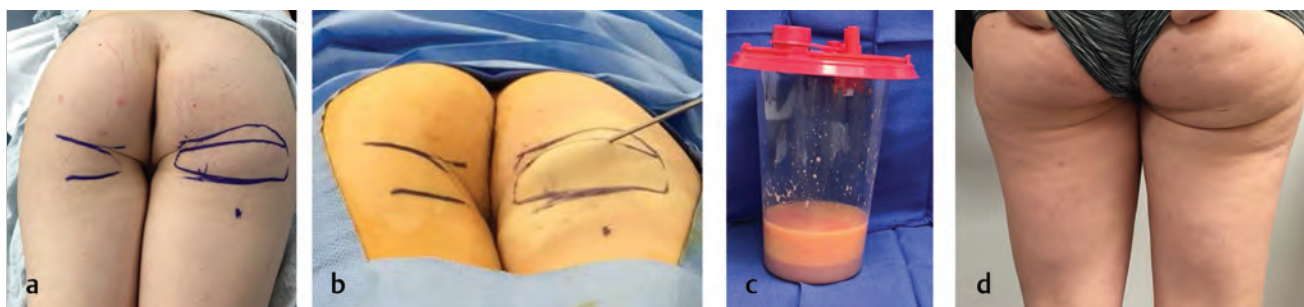


Fig. 29.6 An adolescent female suffered a hematoma following a car accident several years prior to her presentation. She was unhappy with the residual fullness of her right gluteal area. (a) Preoperative appearance with markings of normal gluteal creases on left and elliptical area of soft-tissue overgrowth on right. (b) Intraoperative view of suction-assisted tissue removal. (c) Lipoaspirate. (d) Improved contour 3 months postoperatively.

of blood under the skin flaps which can cause a “second hit” and necrosis of the skin edges.

29.6 Conclusion

The posterior trunk is a favorable place for wounds because there is a significant amount of local tissue that can be mobilized to close defects. Also, scar contracture rarely inhibits function. Usually wounds can be managed low on the reconstructive ladder. Pressure ulcers and other skin defects can be allowed to heal secondarily. If operative treatment is indicated, wide skin undermining and linear closure can close most defects. Paraspinous turnover fascial flaps can be used to cover exposed dura, obviating the need to defunctionalize gluteal or latissimus muscles. Because patients are lying on the incision line, sutures should be maintained until adequate scar strength has been achieved.

29.7 Key Points

- Most defects of the posterior trunk can be managed with local tissues; skin grafts and free flaps are rarely needed.

- Turnover paraspinal fascial flaps should be used to cover exposed dura prior to skin closure.
- Wounds over the spine are repaired with a single linear scar if possible.
- Pressure ulcers will heal secondarily with the elimination of pressure and rarely require operative closure.

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30 Genital Anomalies

Sami H. Tuffaha and Richard J. Redett

Summary

Congenital anomalies of the genitalia include the following: micropenis, aphallia, ambiguous genitalia, hypospadias, epispadias, cloacal exstrophy, transverse/longitudinal vaginal septum, and vaginal agenesis. Patients typically require multidisciplinary care, often with a urologist. Treatment is based on the type and severity of the deformity.

Keywords: genital, anomaly, cloacal exstrophy, pediatric, reconstruction

30.1 Introduction

Genital anomalies can result in debilitating functional and psychosocial sequelae if not treated properly. Beyond the obvious impact on urinary and sexual functioning, a genital defect can damage a child's developing sense of identity and self-esteem, with significant effects on global well-being and interpersonal relationships. As such, reconstructive surgery can have a profoundly positive impact when integrated into a holistic team approach that addresses the urologic, sexual, and psychological pathologies associated with genital defects.

Genital anomalies affecting the penis are more common and more challenging to reconstruct than those affecting the vagina, and will therefore be the focus of this chapter. Penile defects requiring surgical correction in the pediatric population most often result from congenital anomalies, the most common of which are micropenis, aphallia, ambiguous genitalia, epispadias, and bladder and cloacal exstrophy (► Table 30.1). Each of these anomalies is associated with a unique set of pelvic and urologic deformities and physiologic derangements that must be addressed. Understanding how each of these malformations affects normal anatomy and sexual and urologic function facilitates optimal surgical decision-making and maximizes the likelihood of positive outcomes.

30.2 Diagnosis

Diagnosis of congenital penile anomalies typically relies on physical examination. However, deformities are often noted on ultrasound in utero; early recognition allows for prenatal counseling and can help prepare parents for what is to come. Genetic testing is often performed to confirm genetic sex and assess for underlying genetic disorders.

Aphallia is a rare condition in which the penis fails to develop in utero, resulting in complete absence of the penis. Many of these patients have associated genitourinary anomalies that must also be addressed.

The term *micropenis*, or *microphallus*, refers to a normally proportioned penis that is shortened in length (► Fig. 30.1). This condition typically results from insufficient androgen stimulation in utero. In hypogonadotropic hypogonadism, diminished androgen produced is due to insufficient GnRH stimulus from the hypothalamus. In contrast, hypergonadotropic

hypogonadism describes a situation in which there is inadequate androgen release despite normal GnRH stimulus. To diagnose micropenis, the penile length must be more than 2.5 standard deviations below the age-adjusted mean.

The term *ambiguous genitalia* describes a broad set of congenital defects in which the external genitalia have both male and female characteristics. Micropenis is often grouped into this category. Ambiguous genitalia can occur in genetic males or females, and genetic testing is therefore important. In genetic males, ambiguous genitalia can exhibit an array of features, including a small penis resembling a clitoris, a urethral opening above or below the penis, an undifferentiated scrotum resembling labia, and undescended testes (► Fig. 30.2).

Epispadias, *bladder exstrophy*, and *cloacal exstrophy* are thought to belong to a spectrum of genital malformations termed the *exstrophy-epispadias complex (EEC)*. EEC results from failure of mesodermal migration into the cloacal membrane that separates the ectoderm and endoderm of the anterior abdominal wall. This presumably results in premature rupture of the cloacal membrane. The timing and location of rupture dictate whether the patient will develop epispadias, bladder exstrophy, or cloacal exstrophy. Bladder exstrophy presents with a lower abdominal wall defect and severe pubic symphysis diastasis, as well as an exposed, open bladder and urethra and dorsal urethral opening (► Fig. 30.3). Cloacal exstrophy is more severe, with a bilobed bladder separated by the cecum (► Fig. 30.4). Epispadias is the mildest form of EEC and presents with a closed bladder, dorsally open urethral meatus, and mild pubic diastasis.

30.3 Nonoperative Management

Congenital penile deficiency resulting from fetal testosterone deficiency may respond to testosterone treatment, delivered systemically and/or locally. Response to testosterone treatment can be seen in infancy, as well as later in childhood and adolescence. As such, when congenital hypogonadic micropenis is suspected, testosterone treatment should be considered first-line. If medical treatment fails, penile lengthening or phalloplasty can be considered.

Other than for congenital micropenis resulting from fetal testosterone deficiency, medical treatment for congenital penile defects are lacking and surgical intervention is typically warranted.

30.4 Operative Management

30.4.1 General Considerations

The preoperative assessment of a patient with a congenital penile defect begins with careful physical examination and discussion with the referring urologist to determine the extent of the defect. During this initial evaluation, it is important to consider the extent of surgery necessary to achieve an acceptable result. Less severe genital defects can sometimes be addressed by

Table 30.1 Common congenital genital anomalies

Condition	Definition	Etiology	Incidence
Penile anomalies			
Micropenis	Normal proportions but <2.5 standard deviations below appropriate size for age.	Insufficient androgen stimulation in utero with defect in hypothalamic-pituitary-gonadal axis	1:200
Hypospadias	Urethral malformation with ventral termination, often with ventral soft-tissue defect and chordee.	Unclear	1:250
Ambiguous genitalia	Appearance of genitals not clearly male or female.	Congenital adrenal hyperplasia Leydig cell failure Testosterone biosynthesis enzyme defects Androgen insensitivity syndrome Congenital anorchia Gonadal dysgenesis Klinefelter syndrome (47,XXY)	1:4,200
Epispadias	Urethral malformation with dorsal termination, often presenting with small bifid penis.	Failure of mesodermal migration with premature rupture of cloacal membrane. Failure of midline fusion of genital tubercles.	1:120,000
Bladder exstrophy	Abdominal wall defect, pelvic diastasis, penile defect, epispadias, bladder open and exposed	Failure of mesodermal migration with premature rupture of cloacal membrane. Failure of midline abdominal wall fusion after separation of genitourinary and gastrointestinal tract.	1:10,000–1:50,000
Cloacal exstrophy	Abdominal wall defect with extrusion of hind gut, cecum and split bladder, penis and scrotum	Failure of mesodermal migration with premature rupture of cloacal membrane. Failure of midline abdominal wall fusion before separation of genitourinary and gastrointestinal tract.	1:200,000–1:400,000
Aphallia	Agenesis of penis	Failure of the cloacal folds to generate genital tubercle	1:10,000,000
Vaginal anomalies			
Transverse vaginal septum	Tissue bridge across vaginal vault	Improper fusion of Mullerian ducts to urogenital sinus	1:30,000–1:80,000
Longitudinal vaginal septum	Longitudinal tissue bridge dividing vaginal vault	Incomplete fusion of lower Mullerian ducts	1:10,000
Vaginal agenesis	Absence of vagina	Unclear	1:10,000

primary repair, local tissue rearrangement, and/or lengthening procedures. Situations arise in which a defect is otherwise amenable to local flap reconstruction, but the surrounding tissues have been scarred by multiple previous operations. In such cases, tissue expansion has shown great utility. Tissue expansion can also be useful in conjunction with lengthening procedures and repair of hypospadias (► Fig. 30.5). However, when a satisfactory result cannot be achieved with less complex procedures, phalloplasty to reconstruct the entire penis with pedicled locoregional flaps or free tissue transfer can be considered.

The genital defects associated with EEC can pose a difficult reconstructive challenge. Management often involves staged reconstruction of the bladder, urethra, abdominal wall, and genitalia, and requires close coordination and cooperation between the reconstructive surgeon and urologist. The genital

defects associated with EEC vary in severity. In isolated epispadias, the urethral meatus is located on the dorsal surface of the penis at the penopubic angle and the glans is open dorsally. The phallus is typically shortened with dorsal chordee. Epispadias can sometimes be managed with standard lengthening procedures; however, phalloplasty can be used to achieve favorable results in severe cases. In cases of exstrophy, the penis can be halved and separated from the midline. In such cases of severe deformity, phalloplasty is indicated.

In the past, aphallia and severe congenital penile defects were treated with gender reassignment, with bilateral orchiectomy, penectomy (when a penis was present), labioplasty, clitoroplasty, and vaginoplasty. The realization that these patients typically maintain a male identity following surgery, as well as the emergence of effective techniques for phalloplastic reconstruction, has resulted in this practice falling out of favor.

30.4.2 Phalloplasty

The goals of phalloplastic reconstruction include formation of an aesthetically acceptable neophallus, a competent urethra



Fig. 30.1 An adolescent male with micropenis.

allowing for voiding while standing in those patients who do not have continent urinary stomas, enough stiffness to allow for sexual penetration, erogenous and protective sensation, and minimal donor site morbidity. Over the past century, the surgical approach to total phalloplasty has undergone significant evolution. A number of approaches have emerged, each of which provides some, but not all, of the ideal characteristics of a neophallus. The earliest techniques utilized pedicled flaps from the abdomen and groin. More recently, with the advent of microsurgical technique, a number of options involving free tissue transfer have gained popularity. Because they typically provide better function, sensation, and aesthetic outcomes, free flaps have largely replaced pedicled options for phalloplasty.

The *pedicled abdominal flap* was the first type of flap used for total penile reconstruction. It is essentially a pedicled, tubularized rectus abdominus myocutaneous flap based on the deep inferior epigastric vessels. An implant is typically inserted to allow for sexual function. Although a reasonable option when microsurgical capabilities are lacking, this flap lacks sensitivity and the aesthetics are often poor.

The *pedicled groin flap* is an axial flap based on the superficial circumflex iliac vessels, allowing for single-stage reconstruction, with or without an implantable device to provide stiffness. Iliac crest can also be incorporated. Like the pedicled abdominal flap, the pedicled groin flap is limited by poor sensation and aesthetics. Furthermore, it has been noted that the groin flap often results in a small neophallus because of the limited tissue available. Given these limitations, the groin flap for phalloplasty has largely fallen out of favor.

The radial forearm free flap (RFFF) has gained popularity and is considered by many to be the contemporary gold standard for phalloplasty. The RFFF is well suited for neophallus creation in part because it is relatively thin, supple, and hairless, thereby mimicking penile shaft skin. With additional modifications to define the penile subunits, the RFFF now allows for much improved aesthetic outcomes (► Fig. 30.6). Furthermore, because it receives robust sensory innervation from both the medial antebrachial cutaneous (MABC) and the lateral antebrachial cutaneous (LABC) nerves, the neurotized RFFF can provide



Fig. 30.2 A male patient with ambiguous genitalia. Note micropenis resembling clitoris.



Fig. 30.3 A newborn male with bladder exstrophy. Note short urethral plate, scrotal flattening, and chordee. (Adapted from Gearhart JP, Jeffs RD. The bladder exstrophy-epispadias complex. In: Walsh PC, Retik AB, Vaughan ED, et al, eds. *Campbell's urology*. 7th ed. Philadelphia, PA: WB Saunders;1998:1939–1990.)

excellent erogenous and protective sensation, and patients often report the ability to orgasm (Bluebond-Langner and Redett 2011). In order to achieve rigidity, an implant is often inserted in a second stage. However, use of an implant does introduce a risk of extrusion. An alternative approach to provide rigidity without an implant involves harvesting a portion of the radius within the flap; however, this approach is limited by the nuisance of permanent rigidity and a high rate of bone resorption. In addition to prosthesis extrusion, all the fasciocutaneous flaps are prone to high rates of urinary fistulae and strictures. An additional disadvantage of the RFFF is the conspicuous donor site that carries the stigma of gender reassignment in the transsexual community.

The anterolateral thigh flap (ALTF) is a fasciocutaneous perforator flap based on the descending branch of the lateral circumflex femoral vessels. It can be used free of pedicled flap for phalloplasty (► Fig. 30.7). This flap has a number of potential advantages over the RFFF including a less conspicuous donor site, skin that is potentially less pigmented, and greater soft-tissue bulk (Reiner and Gearhart 2004). However, the skin of the ALTF is often much thicker, less supple, and hairier than that of the RFFF. Furthermore, the ALTF is not a good option for patients with large amounts of subcutaneous fat given the difficulty this poses with tubularizing the flap. Like the RFFF, the ALTF can be neurotized for sensation via the lateral femoral cutaneous nerve of the thigh; however, it appears to provide significantly less erogenous sensation in comparison to the RFFF.

The latissimus dorsi myocutaneous flap (LDMF), based on the thoracodorsal vessels, is a reliable option for phalloplasty that provides considerable soft-tissue bulk. Modifications to the LDMF for neurotization have been described, using ilioinguinal to thoracodorsal nerve coaptation. Although protective sensation has been reported using this technique, the mechanism by which this occurs is unclear given that it involves sensory regeneration into a motor pathway. Approximating the thoracodorsal nerve of the flap to the nerve to the gracilis has also been reported, with patients reportedly able to voluntarily contract the latissimus muscle within the flap so as to achieve penetration without an implant.



Fig. 30.4 A newborn male with cloacal exstrophy. (This image is provided courtesy of Dr. John Gearhart.)

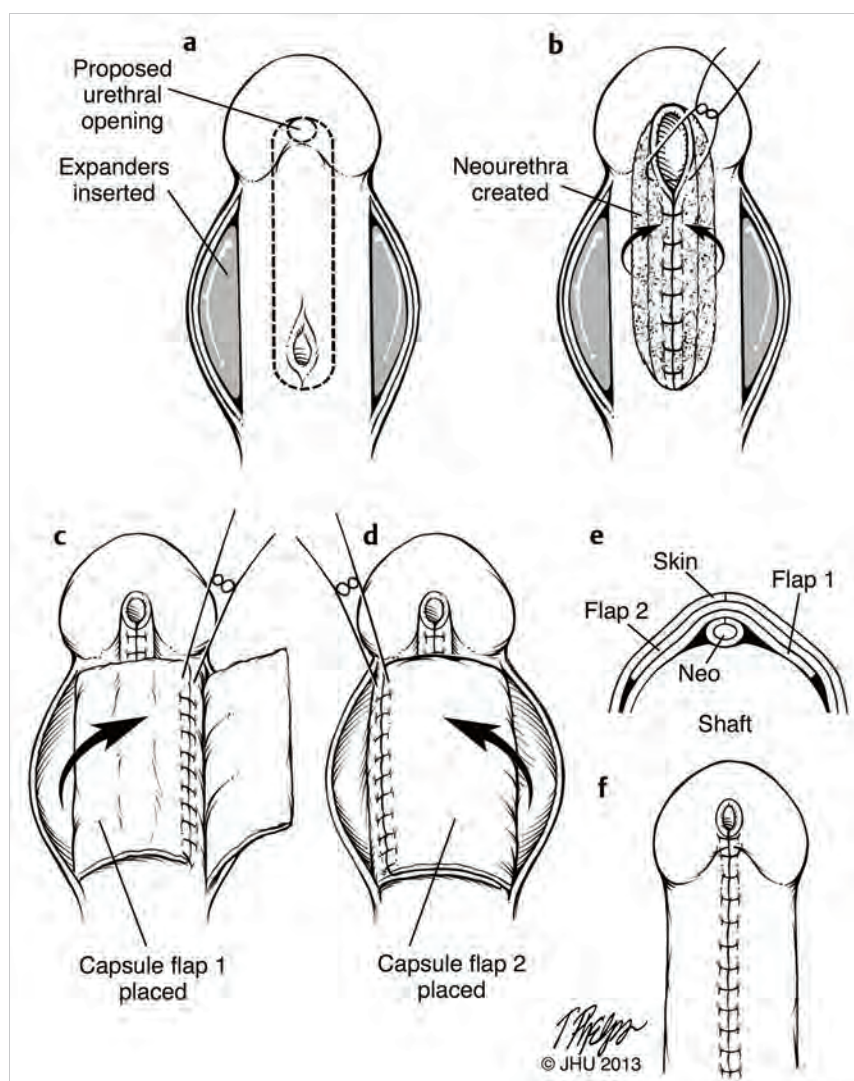


Fig. 30.5 Illustration of hypospadias reconstruction using tissue expansion. (a) Tissue expanders under penile shaft skin. (b–d) The neourethra created using residual urethral plate and expanded shaft skin and covered with fibrous capsule, which formed around tissue expanders. (e,f) Reconstructed phallus with meatus and tip of glans. (Adapted from Rochlin et al. 2014.)

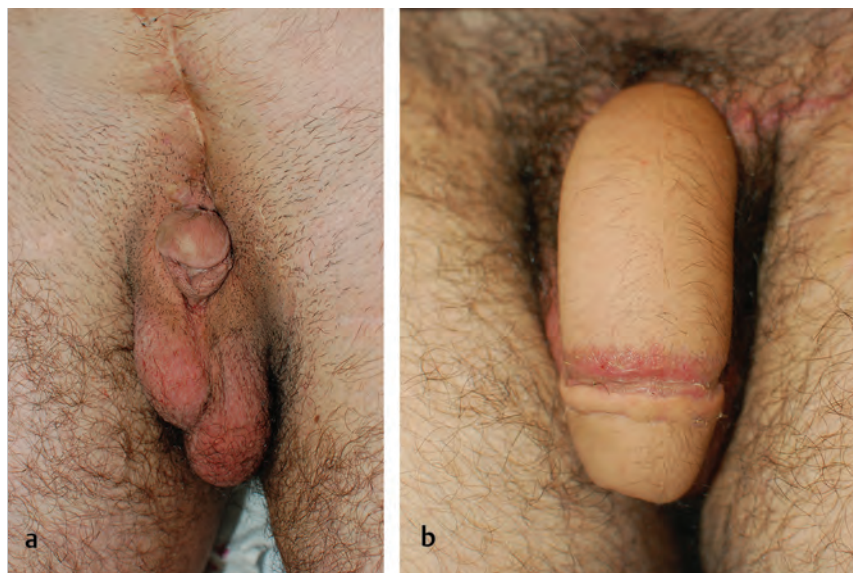


Fig. 30.6 Preoperative photo of male with bladder exstrophy and associated micropenis preoperatively (a) and postoperatively (b) after radial forearm free flap phalloplasty.

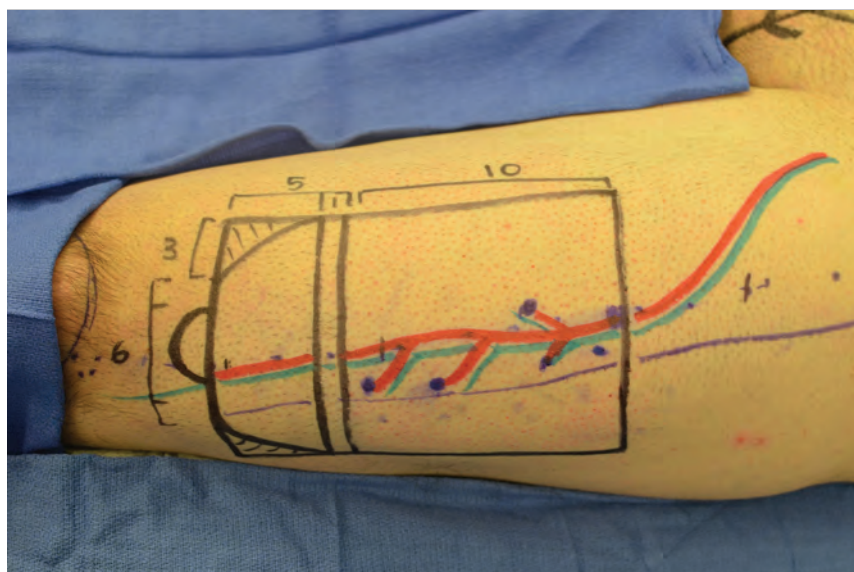


Fig. 30.7 Preoperative markings for pedicled anterolateral (ALT) for phalloplasty.

The fibula osteocutaneous free flap (FOCFF), based on the peroneal vessels, has also been used for phalloplasty. The FOCFF can be neurotized via the lateral sural cutaneous nerve, although the sensibility of the flap is poor in comparison to the RFFF. The primary advantage of the FOCF is inclusion of a portion of the fibula to allow for penetration without an implant. Despite the value of intrinsic rigidity provided by this flap, there are obvious disadvantages to a permanently erect neophallus that can be difficult to conceal and uncomfortable. Furthermore, the bony component within the flap is susceptible to resorption and warping.

30.4.3 Preoperative Planning

Prior to proceeding with phalloplasty, it is important to identify which components of the native penis can be incorporated into a neophallus. If a functional glans is present, it can be incorporated into the ventral base of the neophallus to augment erogenous sensation and provide an ejaculatory duct in those patients with a urinary stoma who are not candidates for creation of a neourethra to the tip of the phallus. When the corpora are present and salvageable, they can also be used for soft-tissue bulk and stiffness within the flap, as well as to help seat an erectile implant. However, incorporation of native components may not be an option, such as in aphallia or other severe malformations. The length and patency of the urethra as well as the status of the more proximal urinary system should also be considered preoperatively. Reconstructive goals should include urination through the neophallus when possible. However, many children with bladder or cloacal exstrophy undergo bladder neck transection and creation of an umbilical continent urinary stoma. In such cases, the native urethra is still brought out through ventral base of the neophallus to allow for ejaculation, but the patient will continue to catheterize the umbilical urinary stoma following reconstruction.

Beyond assessing the genital defect, it is also important to determine whether any associated deficits or comorbidities would preclude or minimize the utility of phalloplastic

reconstruction. It is also important to carefully explain the reconstructive options with the patient's caregivers prior to proceeding. Older children and teenagers should be included in these conversations and should play a central role in decision-making. We generally wait until after puberty to perform total phalloplasty reconstruction. Setting realistic expectations for patients and families is critically important.

30.4.4 Surgical Approach to the Radial Forearm Free Flap for Phalloplasty

The RFFF is considered by many to be the gold standard for phalloplasty and is our preferred method for pediatric patients. To perform the RFFF, the patient is placed in the supine position with the upper extremity from which the flap will be harvested abducted and supinated. A sterile upper extremity tourniquet is used during flap harvest for hemostasis. The flap is marked with dimensions of $14 \times 14 \text{ cm}^2$. This typically includes the entire width and up to three-fourth of the length of the volar forearm, beginning distally at the proximal wrist crease (► Fig. 30.8a and ► Fig. 30.9). Prior to elevating the flap, the neo-glans is defined by raising a transverse split-thickness skin flap across the distal forearm and rolling it under itself where it is secured with resorbable sutures. The donor site of the glans flap is later covered with a full-thickness skin graft to better define the corona and give the penis the appearance of having a circumcision scar (► Fig. 30.8b). This coronal glans flap can also be created once the neophallus is tubed, but we find it easier to raise before the flap is harvested. For patients who require a neourethra, a central longitudinal strip of skin is de-epithelialized, separating the ulnar-sided skin panel (which will line the neourethra) and the radial-sided skin panel (which will provide the external skin of the neophallus).

The RFFF is then raised in a standard fashion from distal to proximal in standard fashion. The radial artery and venae comitantes between flexor carpi radialis and brachioradialis are ligated at the level of the wrist and elevated with the flap from the underlying tendons. Care is given to preserving the paratenon to allow for skin grafting the donor site. The palmaris

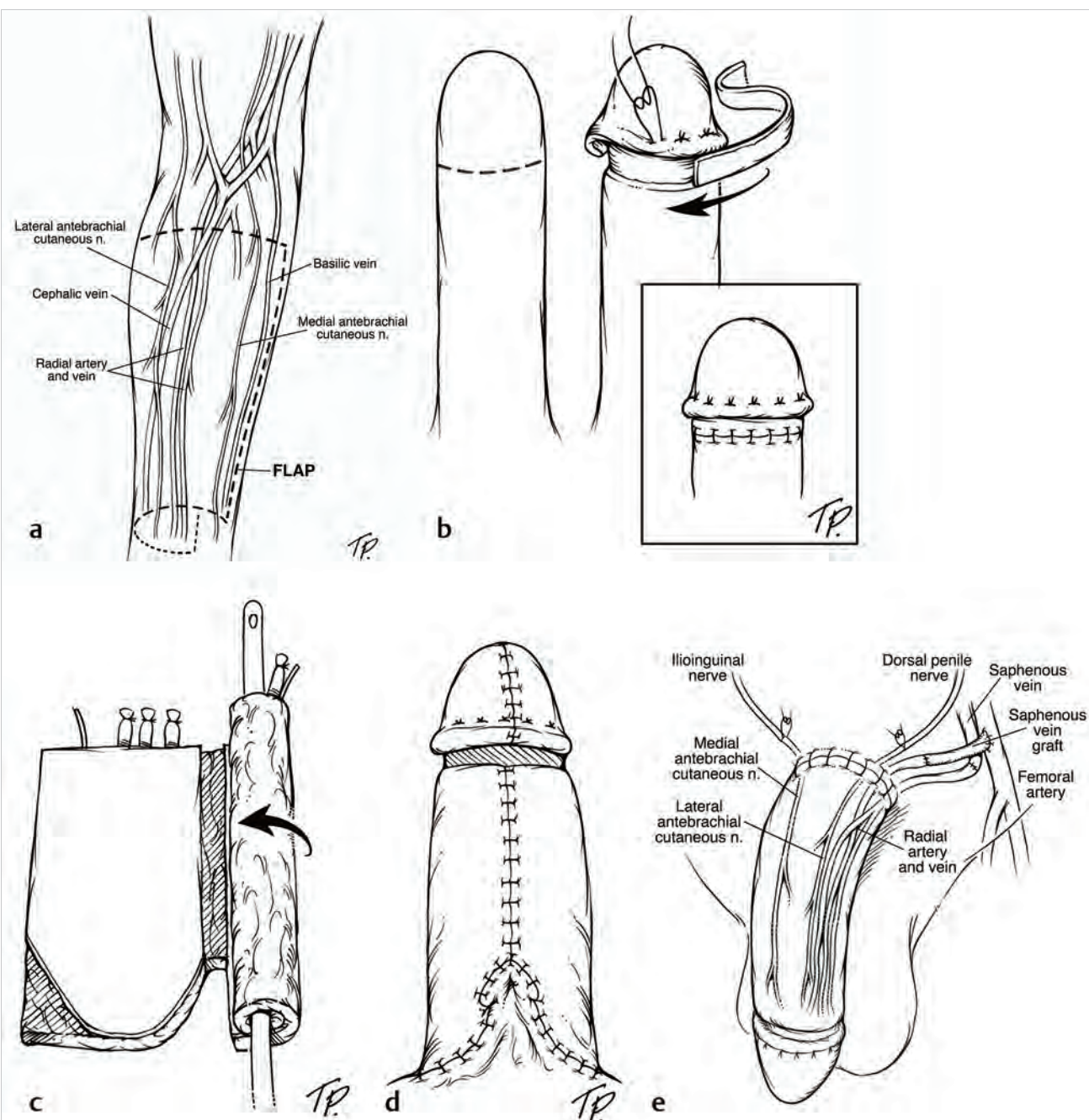


Fig. 30.8 Illustrations of key steps of radial forearm free flap phalloplasty. (a) Flap outlined with important neurovascular structures to be included in flap. (b) Glansplasty and coronoplasty. Skin flap is sutured under itself to create glans. Full-thickness skin graft is applied to donor site of glans flap to create appearance of circumcision scar at corona. (c) Flap being rolled from ulnar to radial. Ulnar skin flap is rolled over itself to create neourethra. Central de-epithelialized skin is buried in flap. Flap is then rolled under itself to externalize the radial skin. (d) Native glans inset into ventral base of neophallus in those patients who are not candidates for urethral reconstruction. (e) Flap transferred showing vascular anastomoses and nerve coaptations.

longus tendon is included in the flap for use in anchoring the neophallus to the pubic bone. The MABC and LABC nerves and the cephalic vein are identified proximal to the flap and harvested with the flap. The flap is tubularized on the forearm before transecting the pedicle (► Fig. 30.10). This involves rolling the flap from ulnar to radial to form the neourethra with the ulnar-sided skin panel. Once the ulnar skin paddle in

internalized, the flap is rolled under itself to externalize the radial-sided skin panel. The de-epithelialized central panel is buried within the flap (► Fig. 30.8c).

Preparation of the recipient site and flap elevation can be performed simultaneously if two surgical teams are available. A pediatric urologist is ideally present to assist with recipient site preparation.

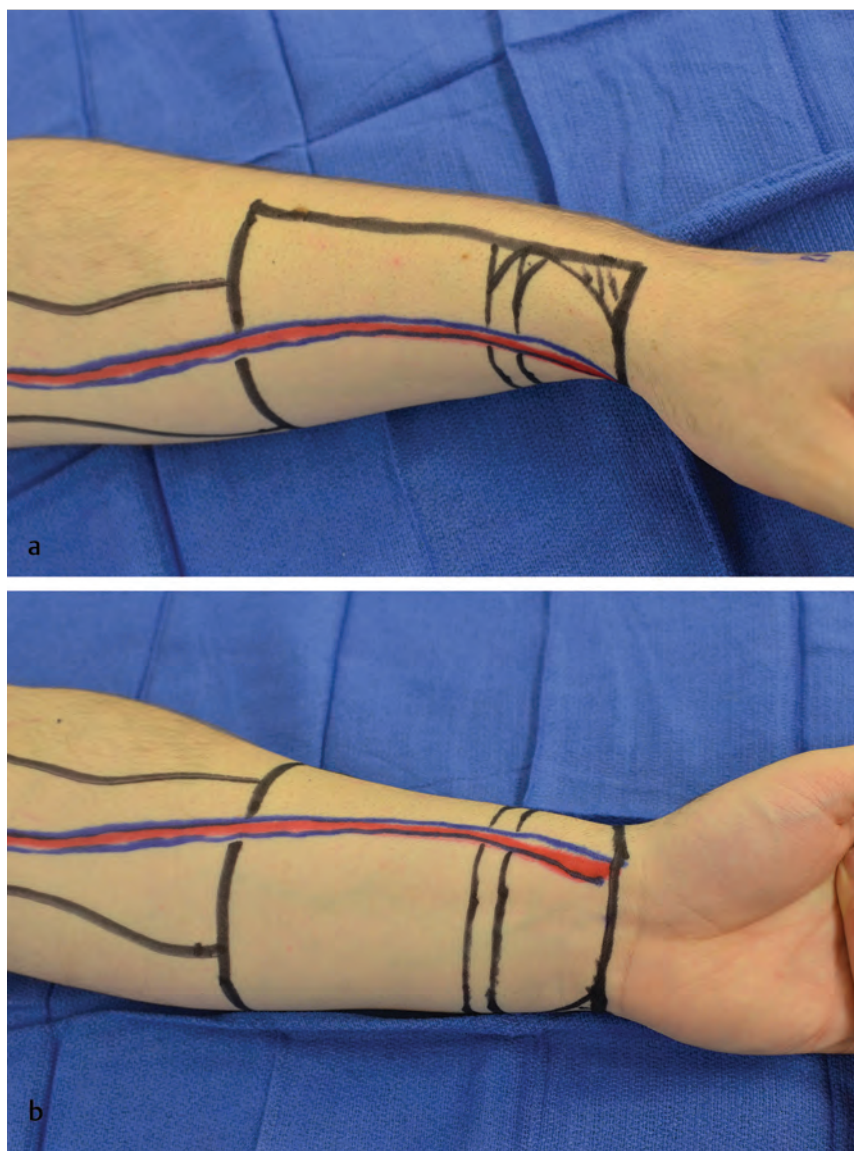


Fig. 30.9 Preoperative markings for radial forearm free flap for phalloplasty.

If the native penis is to be incorporated into the neophallus, the shaft skin is removed and internal structures including urethra and the penile nerves are deconstructed. When a competent urethra in continuity with the proximal urinary tract is present, the corpus spongiosum is severed from the glans so that it can be anastomosed to the neourethra within the flap. However, when the bladder neck has previously been transected and the patient voids from an umbilical stoma, the urethra is left in continuity with the glans, and the external meatus is externalized through the ventral base of the neophallus to allow for ejaculation (► Fig. 30.8d). In either case, the glans is incorporated into the neophallus. To do this, it is de-epithelialized and buried into the base of the neophallus, either at the time of phalloplasty or, more conservatively, at a later stage to account for possible flap failure.

The dorsal penile nerves for erogenous sensation, and either the ilioinguinal or iliohypogastric nerve for protective sensation, are dissected in the pelvis and approximated to the MABC and LABC nerves of the flap. A number of possible recipient site vessels can be anastomosed to the radial artery to perfuse the

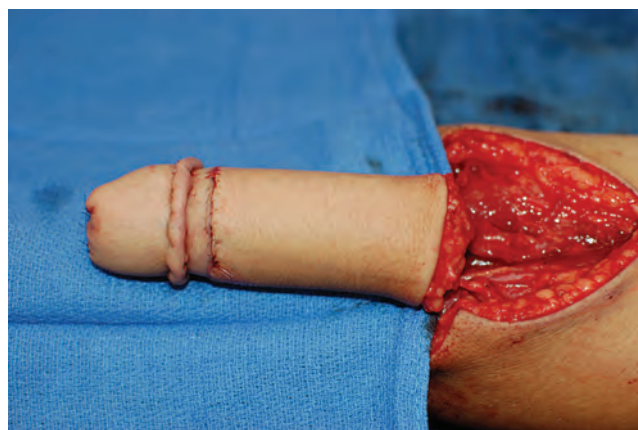


Fig. 30.10 Intraoperative photograph of radial forearm free flap tubularized and prelaminated on forearm prior to transfer.

flap, including the superficial circumflex iliac, inferior epigastric, external pudendal, and femoral arteries. The accompanying veins, as well as the greater saphenous vein, can be used to provide venous outflow via the venae comitantes of the radial artery and the cephalic vein. We perform as many venous anastomoses as possible to help reduce postoperative swelling and venous congestion (► Fig. 30.8E).

The neourethra within the flap is anastomosed to the native urethra over a 10 or 12 French urinary catheter at the time of phalloplasty. Although the senior author's preferred method for neourethra construction is described above, alternative approaches include prelaminating the flap with a full-thickness skin graft or inserting a tubed skin or buccal mucosal graft at a later stage.

Insertion of the penile prosthetic implant is performed at a later stage and is typically delayed until late adolescence or early adulthood to minimize implant-associated risks, including soft-tissue erosion, infection, and extrusion. Prior to implant insertion, protective sensation should be present and the patient should exhibit a high level of maturity. We wait a minimum of 1 year after phalloplasty before inserting the implant to insure the return of protective sensation before the procedure.

30.5 Complications

The RFFF is the most popular option for phalloplasty because it allows for excellent aesthetics as well as consistent erogenous and protective sensation. However, as with all methods of phalloplasty, there are significant limitations to the RFFF that must be given serious consideration. While all of our patients who underwent RFFF phalloplasty report the ability to achieve orgasm and satisfaction with neophallus appearance, we have also noted that roughly half of our patients experience significant complications involving urinary strictures and fistulae and/or implant-associated complications requiring implant explantation; these findings are consistent with other large series.

30.6 Conclusion

There are a number of congenital anomalies affecting the penis, each of which requires a different approach to reconstruction. It is important to understand how the different anomalies affect normal anatomy as well as the various reconstructive options that can be used to correct them. A multidisciplinary team is required to address the urologic, sexual, and psychological sequelae associated with genital anomalies. Although

autogenous phalloplasty can offer favorable results, there are a number of drawbacks including donor site morbidity and complications involving prosthesis extrusion and urinary strictures and fistulae. Tissue engineering and vascularized composite allotransplantation may offer improved results in the future.

30.7 Key Points

- Congenital anomalies are the most common cause of penile defects requiring reconstruction in the pediatric population.
- Genetic males with aphallia, severe micropenis, or ambiguous genitalia should be considered for phalloplasty; early gender reassignment surgery for these patients has fallen out of favor given they often maintain their male identity.
- Congenital penile anomalies resulting from fetal testosterone deficiency may respond to testosterone therapy.
- Some penile defects can be corrected with tissue expansion, local tissue rearrangement, and lengthening procedures. More severe defects require phalloplasty.
- The goals of phalloplasty include formation of an aesthetically acceptable neophallus, a competent urethra allowing for voiding while standing in those patients who do not have continent urinary stomas, enough stiffness to allow for sexual penetration, erogenous and protective sensation, and minimal donor site morbidity.
- The authors consider the RFFF to be the gold standard for phalloplasty because it offers superior aesthetics and sensation.
- Complications of phalloplasty include donor site morbidity, prosthesis extrusion, and urinary strictures and fistulae.
- Tissue engineering and vascularized composite allotransplantation may offer improved results in the future.

Suggested Readings

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31 Lower Extremity

Arin K. Greene and Amir H. Taghinia

Summary

Common indications for lower extremity reconstruction in children include the following: constriction ring, lymphedema, syndactyly, defects following skin lesion removal, and trauma. Principles of reconstruction are similar to adults. Management is based on the type of deformity.

Keywords: leg, pediatric, constriction ring, lymphedema, syndactyly, nevus, trauma

31.1 Introduction

In the pediatric population, plastic surgical problems can be congenital or acquired. The most common idiopathic disorders managed by plastic surgeons are syndactyly, macrodactyly, constriction rings, and lymphedema. Acquired disorders of the leg frequently arise from trauma or following extirpation of a lesion. Traumatic injuries are managed similarly to adults. However, because of the young age of the patients and long projected lifespan, aesthetic concerns and function are particularly emphasized by patients/families. We favor managing patients as low on the reconstructive ladder as possible.

31.2 Diagnosis

Diagnosis of lower extremity disorders primarily is made by history and physical examination. Traumatic wounds are evaluated for tissue loss, nerve injury, fractures, and foreign bodies. Usually adequate local tissue is available to cover bone or hardware proximal to the knee. Large areas of exposed bone or hardware involving the knee and midtibia area may be covered using regional gastrocnemius or soleus flaps. However, the threshold to use the flaps is higher in children compared to adults because of the long-term potential of reduced function. Significant tissue loss involving the lower one-third of the leg may require free-tissue transfer. Plain radiography is used to rule out fractures and foreign bodies following major traumatic injuries. X-ray also is used to assess syndactyly and macrodactyly to determine an underlying osseous abnormality.

The diagnosis of lymphedema can be difficult. The term “lymphedema” often is used generically to describe any condition that causes overgrowth of an extremity; one-fourth of patients referred to our lymphedema center do not have the disease. Lymphedema is the chronic, progressive swelling of tissue due to inadequate lymphatic function. In the pediatric population, 97% of cases are due to maldevelopment of the lymphatic system (primary) and 3% are due to inguinal lymphadenectomy/radiation (secondary). Primary lymphedema affects 1/100,000 persons and almost always affects the lower extremity; the genitalia is the second most common site. Males typically exhibit bilateral swelling during infancy, while girls usually present with unilateral edema in puberty.

Lymphedema almost always affects the distal extremity and then progresses proximally. If the hand or foot is not involved,

the patient is unlikely to have lymphedema. Over time, subcutaneous lymph stimulates adipose deposition and fibrosis, which causes nonpitting edema and a positive Stemmer sign (the inability to grasp the base of the second toe or finger). Lymphedema is usually painless and does not cause ulceration. Definitive diagnosis of lymphedema requires lymphoscintigraphy; almost all patients referred to our center undergo this imaging study.

31.3 Nonoperative Treatment

Congenital anomalies do not require operative intervention unless they are causing a functional or “cosmetic” problem. Because long-term memory and self-esteem begin to form at approximately 4 years of age, elective correction of deformities not causing a functional problem can be performed at 3 years of age. Alternatively, patients can be observed until they are old enough to request improvement of a deformity (e.g., syndactyly, macrodactyly, benign skin lesion).

Most patients with lymphedema do not require operative intervention. Lymphedema is associated with a high risk for infection; therefore, patients are counseled about washing and moisturizing the affected area regularly to prevent desiccation, skin opening, and cellulitis. Patients are encouraged to participate in all physical activities, including exercise. The first-line therapy for lymphedema is compression garments, which reduce extremity volume and minimizes progression. Pneumatic compression devices provide intermittent pressure via an inflatable sleeve. Pneumatic compression therapy is efficacious, simple, and convenient because it is performed at home and is not dependent on a therapist. Complex decongestive therapy employs skin care, manual lymphatic drainage, compression bandaging, and exercise. We generally do not recommend this intervention because it is particularly difficult in the pediatric population and requires a substantial time commitment for the patient/family and the reliance on a provider.

31.4 Operative Treatment

31.4.1 Constriction Ring

Constriction ring syndrome (CRS) can affect any portion of the lower extremity, though it presents more commonly in the toes. There is a spectrum of involvement, from superficial scarring to deep bands down to bone resulting in functional detriment, growth disturbance, joint deformity (e.g., club foot), and/or distal lymphedema. The most severe manifestation can be congenital amputation at any level.

Operative treatment of CRS should address the functional and aesthetic issues of the affected limb. Collaboration with an orthopaedic colleague is vital in cases of joint deformity (e.g., club foot) or where there is significant restriction of joint motion. For bands affecting the legs or thighs, excision and flap advancement are the ideal treatment. Most advocate treatment prior to ambulation to take advantage of the extra soft tissue of

an infant and easier immobilization. The authors employ the technique advocated by Upton. An assessment of tissue laxity and movement is performed. Bands that are deep and narrow are ideal as they provide plenty of soft tissue to restore contour. The ring is then marked and the skin is excised. Separate adipofascial and skin flaps are raised. The adipofascial flaps are mobilized and closed and then the skin flaps are approximated ideally away from the line of closure of the adipofascial flaps. This technique provides the best contour. Z-plasties are advocated extensively in the literature but are rarely needed. If the ring is deep or if there are multiple closely spaced rings, then a staged approach may be prudent (► Fig. 31.1).

31.4.2 Lymphedema

A patient must fail conservative therapy and have significant morbidity before a surgical procedure is considered (e.g.,

psychosocial distress, recurrent infections, decreased function). Suction-assisted lipectomy is our preferred operative technique for extremity lymphedema because it gives consistently favorable results with minimal morbidity. The technique removes the abnormally hypertrophied subcutaneous adipose tissue and reduces excess extremity volume by approximately 75% (► Fig. 31.2). Liposuction does not cure lymphedema, and patients must continue their compression regimen. Recurrence has not been noted with 15 years of follow-up. Microsurgical procedures have been performed using lymphatic-venous anastomosis (LVA) and vascularized lymph node transfer. These procedures theoretically might have efficacy in mild lymphedema, before fibroadipose deposition has occurred. However, patients with mild lymphedema rarely have significant morbidity and can be managed well with compression. Because patients with primary lymphedema have hypoplastic or absent lymphatics, they are poor candidates for microsurgical procedures. LVA and



Fig. 31.1 A young child with a constriction ring of the lower extremity. (a,b) Preoperative appearance. (c,d) After circumferential excision, advancement of cephalad and caudad fasciocutaneous flaps, and skin closure.



Fig. 31.2 A female with adolescent-onset primary lymphedema of the left lower extremity. (a) Preoperative appearance. (b) Intraoperative view of subcutaneous adipose removal using suction-assisted lipectomy. (c) Lipoaspirate. (d) Postoperative result.

lymph node transfer have not shown consistent improvement in lymph flow and reduction in extremity volume. Lymph node transfer has been associated with donor site lymphedema following node harvest. Because patients with primary lymphedema are at risk of developing lymphedema in their other limbs, they are at highest risk for developing donor site lymphedema following removal of lymph nodes.

31.4.3 Syndactyly

Syndactyly most commonly affects the second webspace in the foot. In Apert syndrome, the syndactyly affects all four webspaces but rarely is there bony coalition distally. In most patients, syndactyly of the foot does not cause major functional problems. Most children and adults are able to walk, run, and participate in sports. Treatment is usually requested by parents in early infancy or by patients during teenage years as they become more self-conscious about their appearance.

Under general anesthesia and tourniquet control, dorsal and plantar triangular flaps are planned for commissure reconstruction (► Fig. 31.3). Plantar zigzag incisions, similar to those used for release of hand syndactyly, are not needed. Instead, straight-line releases are used and these rarely result in contractures. The resulting defects are covered with full-thickness skin grafts from the lower abdomen. Three weeks of cast immobilization and non-weight bearing is critical to allow the skin grafts to fully take. The ideal time to perform the operation is in infancy just before the patient begins to ambulate.

31.4.4 Reconstruction Following Skin Lesion Removal

The lower extremity is an unfavorable location to remove large lesions of the integument because (1) skin redundancy is minimal, (2) ambulation places tension on the incision line, and (3) gravity favors swelling, which increases stress on the incision line. Options to reconstruct cutaneous defects following extirpation include (1) allowing it to heal by secondary intention, (2) linear closure following skin undermining, (3) placement of a skin graft, (4) tissue expansion, and (5) regional or free tissue transfer. In extremities, one should consider cast or removable knee immobilization after lesion excision to ensure better healing and avoid dehiscence.

Timing of intervention falls into three categories: (1) 6 to 12 months of age, (2) 3 to 4 years of age, and (3) late childhood/early adolescence. It is best to perform elective procedures after 6 months of age because the infant's physiology approximates that of an adult, which reduces the risk of anesthesia. Because most infants begin ambulating around 12 months of age, it is preferable to remove lesions before this time to reduce the risk of suture line dehiscence. If patients present after 12 months of age with lesions causing a deformity only (e.g., infantile hemangioma, benign pigmented nevus), then removing the lesion before 4 years of age will eliminate the deformity before the child's long-term memory and self-esteem begins to form. Some parents will prefer to wait until a child is old enough to participate in the decision to extirpate a lesion, which can occur in late childhood or early adolescence.



Fig. 31.3 Toe syndactyly treated with dorsal and plantar triangular flaps. (a) Preoperative appearance. (b,c) Markings of the flaps that are raised sharply in the subdermal plane. Once the syndactyly has been released, the flaps are transposed into the commissure. (d,e) Full-thickness skin grafts from the lower abdomen are used to resurface the skin defects on each side of the web.



Fig. 31.4 A young child with an osseous lesion of the toe. **(a)** Preoperative view. **(b)** After resection. The wound was allowed to heal secondarily to avoid complicated reconstruction. The child was allowed to immediately ambulate and wash the area. **(c)** Favorable postoperative appearance.

Small defects or areas on the foot that would necessitate complicated reconstruction can be allowed to heal secondarily (► Fig. 31.4). Allowing the wound to heal by contraction and epithelialization prevents the risk of infection and suture line dehiscence. In addition, it facilitates recovery for the patient who can immediately resume full activity and bathe. Almost all skin lesions can be removed by lenticular excision and linear closure (► Fig. 31.5); large areas are managed by serial excision (► Fig. 31.6). We prefer this procedure over skin grafting because grafts leave a large circumferential scar as well as an indented, hairless area. Many patients are unhappy with the appearance of a skin graft and request that it be removed. We

believe serial excision is superior to tissue expansion because it is associated with less morbidity and the child ultimately has a single linear incision instead of multiple scars from rotation/advancement of expanded skin flaps. Serial excision expands the integument without the use of expanders; the technique involves short outpatient procedures with minimal morbidity and does not require outpatient injections of saline. External tissue closure systems can aid the closure of large wounds. Occasionally, large circumferential lesions require skin grafting or tissue expansion because serial excision can only be performed if normal skin is present on either side of the lesion (► Fig. 31.7).



Fig. 31.5 Immunocompromised child with a squamous cell carcinoma of the leg. **(a)** Preoperative appearance. **(b)** Linear closure following extirpation with wide margins.

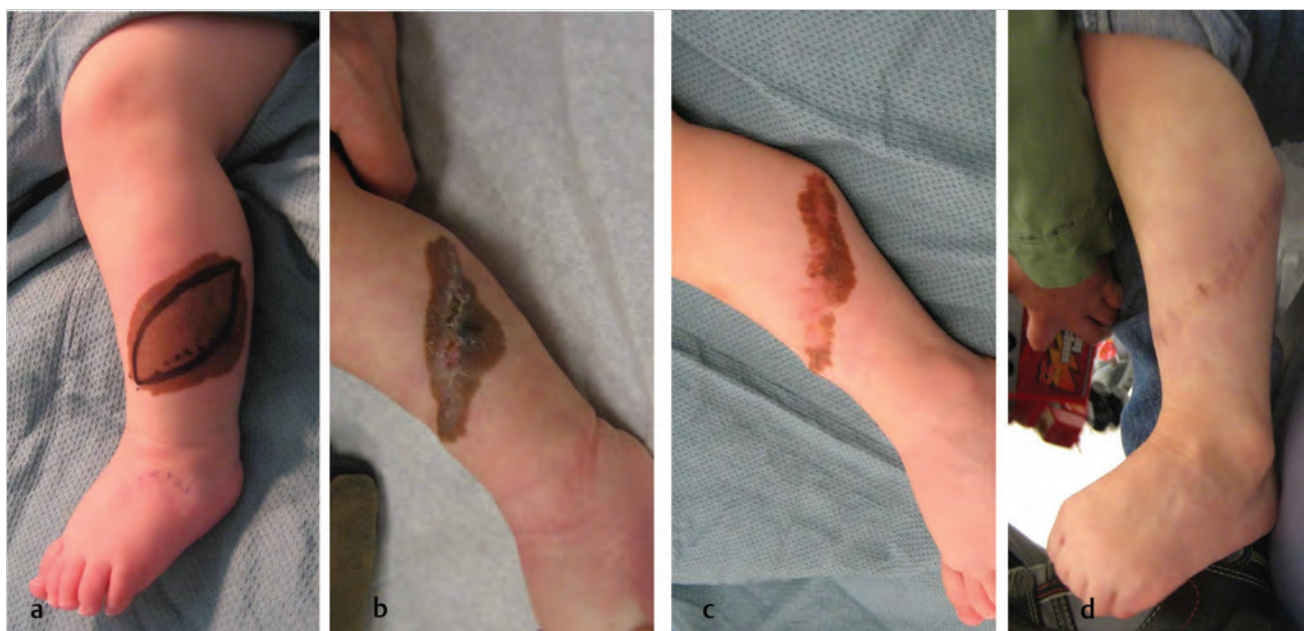


Fig. 31.6 Removal of a large congenital skin lesion. (a) Preoperative appearance of the melanocytic nevus. Outline illustrates the initial area to be resected. (b) After first-stage resection. (c) Following second-stage serial excision. (d) Final appearance after third-stage removal.

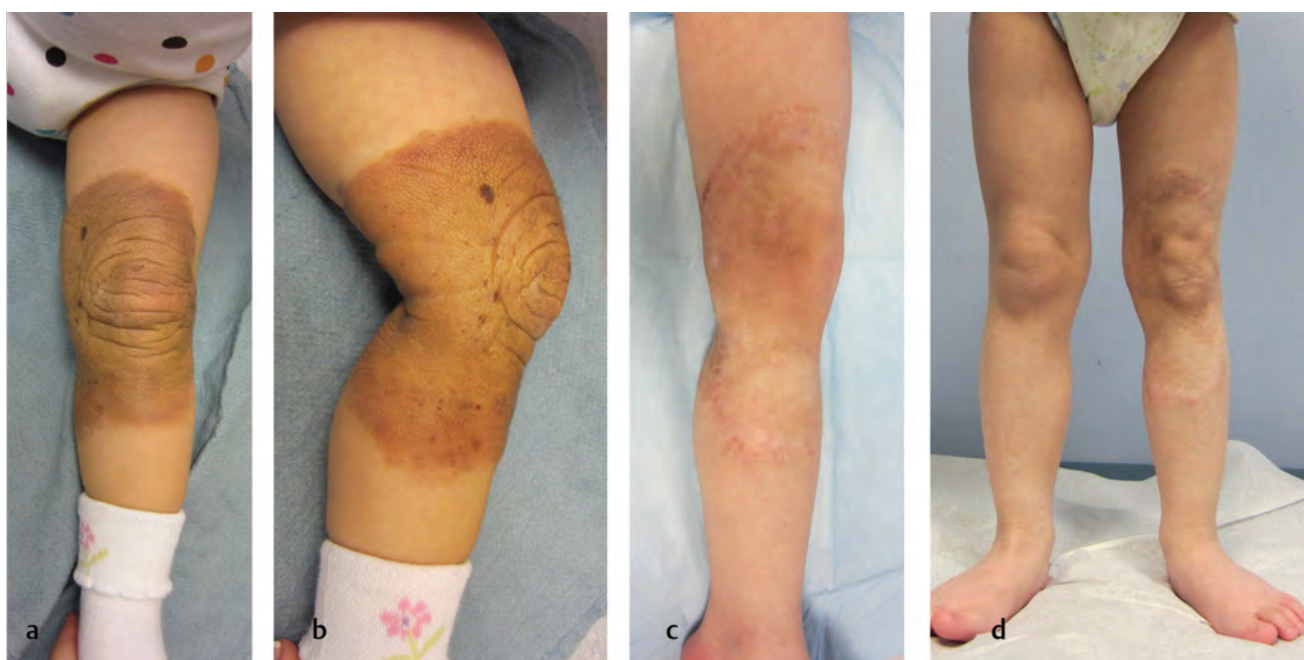


Fig. 31.7 Giant pigmented nevus of the lower extremity. (a,b) Preoperative appearance. Because the lesion was large and circumferential, it was not amenable to serial excision. The patient underwent tissue expansion of the trunk to obtain adequate full-thickness skin to cover the area following extirpation. (c,d) Postoperative result after resection and application of a full-thickness skin graft.

31.4.5 Trauma

Reconstruction of traumatic defects is based on the size, location, and depth of the wound (► Fig. 31.8). Superficial areas can be allowed to heal secondarily (with or without vacuum-assisted wound closure). Larger defects may be closed with delayed primary closure (with or without external tissue expansion; ► Fig. 31.9, ► Fig. 31.10). Skin grafts are only necessary if the wound cannot be closed linearly by advancing skin

flaps (► Fig. 31.11). Skin grafts should be avoided if possible because they cause a worse deformity than a linear scar, and patients often will request that the graft be removed secondarily.

Deep wounds with exposed bone can be allowed to heal secondarily (if the area is small), or closed with approximation of adjacent tissues. Localized areas of exposed bone or tendon can be managed with vacuum-assisted wound closure to decrease the area of the wound and generate granulation tissue



Fig. 31.8 Penetrating trauma causing transection of the tibial nerve. (a) Preoperative appearance. (b) Intraoperative view of the divided nerve. (c) Following nerve repair. (d,e) Good function postoperatively.

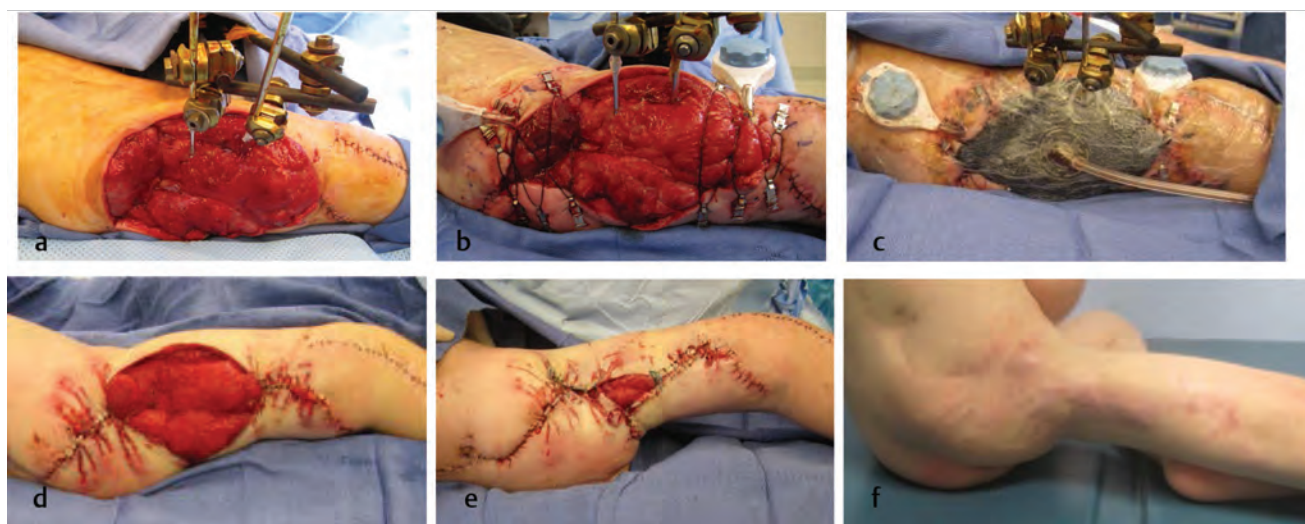


Fig. 31.9 Traumatic lower extremity wound with significant skin loss. (a) Preoperative appearance with external fixator in place. (b) Application of external tissue expander. (c) Negative pressure wound therapy over closure device. (d) Serial delayed primary closure after removal of the wound closure system and continued vacuum-assisted wound closure therapy. (e) Small wound after completion of serial delayed primary closure and negative pressure wound therapy. The area was left open to heal secondarily. (f) Closed wound with a linear scar.



Fig. 31.10 Traumatic lower extremity wound managed with delayed serial closure, external tissue expander, negative pressure wound therapy, and a split-thickness skin graft. (a) Preoperative appearance. (b) Application of external wound closure device and vacuum-assisted sponge. (c) Most of the wound was able to be closed with delayed primary closure giving a single linear scar. (d) The area involving the knee was unable to be closed linearly and underwent reconstruction with a split-thickness skin graft.

amenable to skin grafting. We do not favor alloplastic substance (e.g., integra, dermal substitutes) placed over bone or tendon because vacuum-assisted wound closure will provide adequate granulation to simplify reconstruction. Large areas of exposed bone, tendon, or hardware in the lower leg require regional muscle flap closure (e.g., gastrocnemius, soleus) or free tissue transfer (► Fig. 31.12, ► Fig. 31.13, ► Fig. 31.14).

31.5 Complications

The most common complication following a procedure on the lower extremity is suture line dehiscence. The larger and more distal the skin defect, the greater the risk of wound breakdown. Suture line dehiscence can be minimized using several strategies. First, sutures can be left for at least 4 weeks, when the wound has achieved 40% of its strength. Second, the patient is placed in a knee immobilizer for 2 to 3 weeks to limit stress on the incision line. The knee immobilizer can be removed when the patient is sleeping. To help prevent the scar from widening after removing the sutures, it can be taped with steri-strips 6 weeks postoperatively when the scar has achieved its full strength.

31.6 Conclusion

Lower extremity reconstructive problems in children are different than those in adults. Several congenital disorders that are not managed in the adult population exist. Principles of extirpation of lesions and reconstruction can be similar to adults. However, children are less likely to follow postoperative instructions and tolerate suture removal. Often, immobilization of the extremity should be performed after an operation. The pediatric population does not have arterial or venous disease and thus can best tolerate wide skin undermining and closure of wounds linearly. In general, children should be managed as low on the reconstructive ladder as possible.

31.7 Key Points

- Most cutaneous lesions and skin defects in the lower extremity can be closed with serial excision/delayed primary closure.
- Skin grafts and subcutaneous tissue expanders should be avoided if possible.
- Maintenance of sutures for several weeks and immobilization of the extremity can reduce the risk of suture line dehiscence.

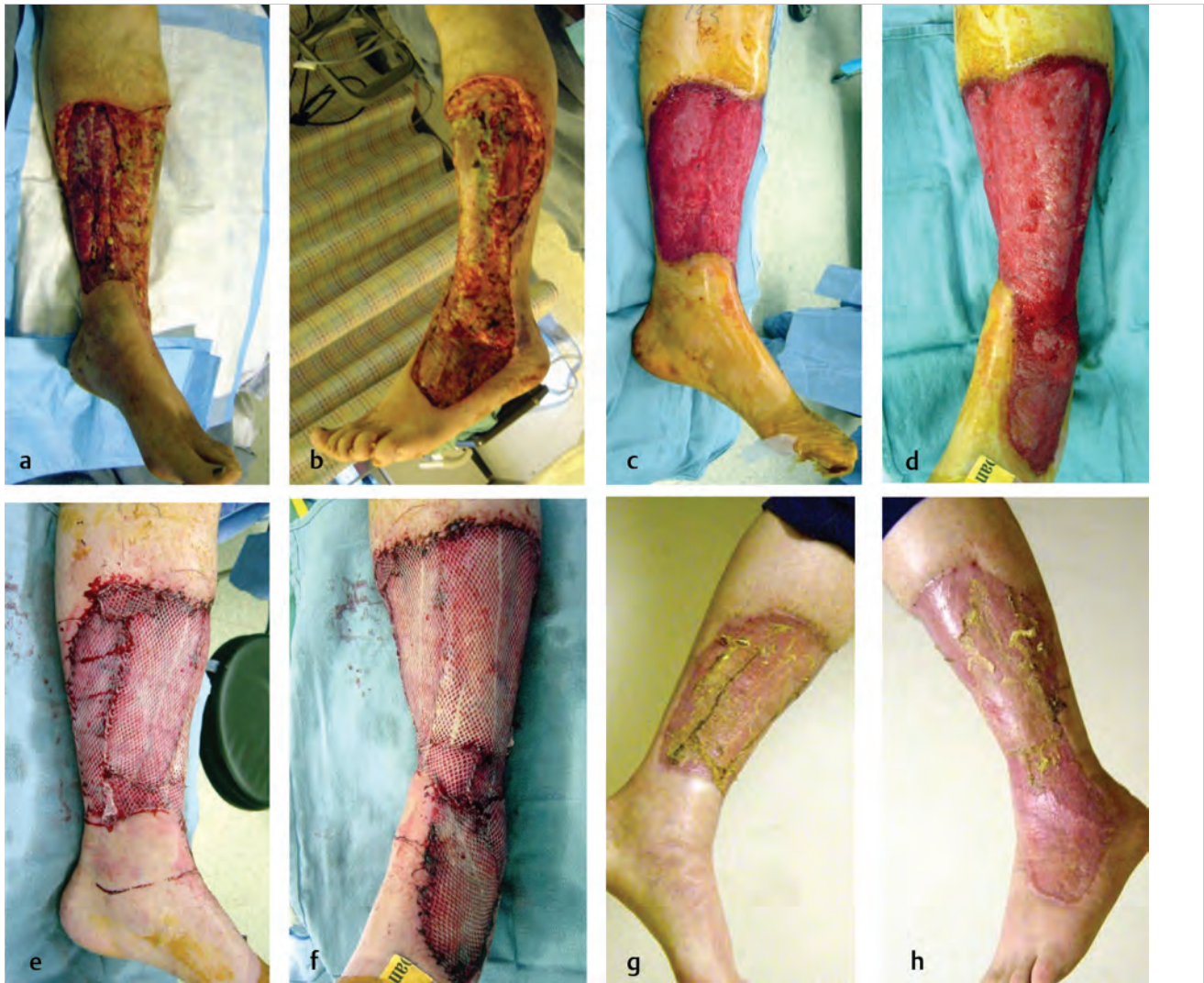


Fig. 31.11 A male with a traumatic injury. Because the skin loss was extensive and circumferential, his wound was managed with a split-thickness skin graft. (a,b) Preoperative wound illustrates exposed tendon and periosteum. (c,d) Healthy granulation tissue over all structures following treatment with negative pressure wound therapy. (e,f) Split-thickness skin graft coverage of wound. (g,h) Postoperative appearance.



Fig. 31.12 Traumatic foot injury with exposed bone managed with a local transposition flap and skin graft. (a) Preoperative appearance. (b) Exposed bone following irrigation and debridement. (c) Local flap used to cover the area of exposed bone. (d) Donor defect is covered with a skin graft.



Fig. 31.13 Exposed hardware after tibial fracture managed with medial gastrocnemius flap and split-thickness skin graft. (a) Preoperative view shows the reconstruction plate. (b) Following coverage of plate with the muscle flap. (c) Skin graft over exposed muscle.

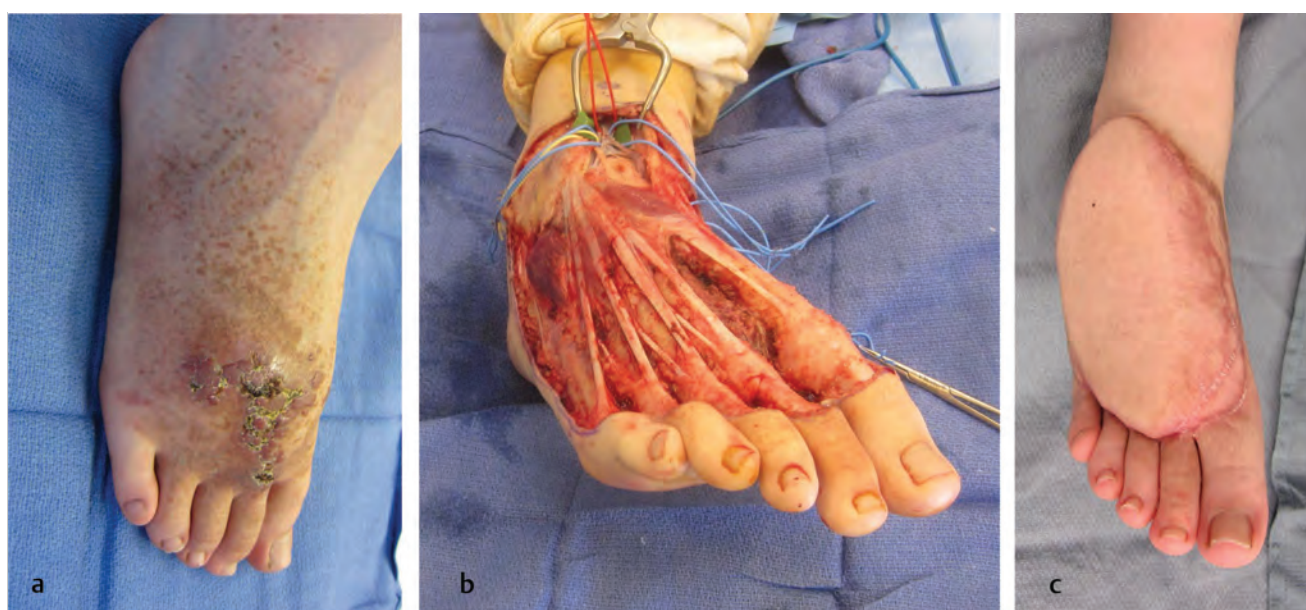


Fig. 31.14 An arteriovenous malformation of the foot was managed with completed extirpation and reconstruction with a free tissue transfer. (a) Preoperative appearance. (b) Following resection. (c) Inset of anterolateral thigh free flap.

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Part V

Upper Extremity

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32 Embryology and Classification of Hand Differences

Bran Sivakumar and Gill Smith

Summary

Congenital hand differences arise from genetic or environmental influences on the fetus during development. Embryogenesis of the upper limb occurs between the fourth and eighth weeks of gestation. Normal limb development requires a time-dependent regulated sequence of transcription factors and growth factors. It is best considered by simplification into the three axes of development: proximodistal, anteroposterior, and dorsoventral. Dysmorphology can result from disruption at any stage during upper limb development and involve any structures. The International Federation of Societies for Surgery of the Hand morphologically based classification of congenital hand differences predated current knowledge of etiology and embryological development. The Oberg–Manske–Tonkin classification attempts to incorporate this to allow clinicians and developmental biologists to communicate in a common language. It offers a framework for conveying the etiology of the anomaly while remaining flexible to new developments.

Keywords: embryology, OMT, Oberg–Manske–Tonkin, hand, limb bud, development, dysmorphology

32.1 Introduction

The incidence of upper limb anomalies is approximately 1 in 450 live births based on United Kingdom data. If one anomaly is identified, there is also an increased likelihood of a further non–upper limb anomaly. There is huge variety in the severity of the appearance and the functional deficit associated with a congenital hand difference with many upper limb malformations resulting in relatively little disability due to adaptive strategies. The range of congenital anomalies is vast, ranging from a barely discernible mild thumb hypoplasia to complete agenesis.

Management of the patient requires accurate assessment and diagnosis, clear communication with the family and child, and may require therapy or surgical intervention. To properly assess such patients, a sound understanding of the pathogenesis of the anomaly, and thus of normal upper limb embryology, is required. In this chapter, we will discuss normal embryology and development of the upper limb. We will then relate this to the stages during which anomalies can develop, and finally the accepted framework for classification of such congenital anomalies.

32.2 Early Embryology

The zygote undergoes cleavage during the first week of development to produce a blastocyst, with an inner embryoblast and an outer trophoblast. In week 2, the dorsoventral (DV) axis of the embryo is established, with splitting of the embryoblast into the primary ectoderm and primary endoderm. In the third week, the primitive streak arises at the caudal end of the bilaminar germ disc, establishing the longitudinal axis of the embryo. The primitive node forms at the cranial end of the primitive streak and connects to the notochord. The notochord

process is a mesodermal midline structure that induces the development of the neural plate in the ectoderm.

Gastrulation begins in the third week. During this process, the definitive endoderm, ectoderm, and mesoderm are formed, converting the embryo into a trilaminar germ disc. The primitive streak regresses, during which mesodermal cells on either side of the notochord form the somatic mesoderm. These later differentiate into the axial skeleton, voluntary musculature, and part of the skin dermis. A less pronounced lateral plate mesoderm forms and splits into a ventral layer associated with the endoderm, which forms the mesothelial covering of the viscera and heart, and a dorsal layer associated with the ectoderm, which forms the body wall lining and dermis.

32.3 Limb Bud Development

Limb bud development occurs between the fourth and eighth weeks of embryonal development, with a later prolonged growth phase after formation. The notochord expresses sonic hedgehog (SHH) between days 21 and 22, which regulates the initiation of the limb bud. The limb bud is an outgrowth of somatic mesoderm and lateral plate mesoderm into the overlying ectoderm, which forms an outer ectodermal “cap.” The upper limb buds are first noted on the lateral body wall 24 days after fertilization in the lower cervical region, at the level of C5–C8. At 26 days, a ridge of bulging ectoderm develops on the ventromedial border of the limb bud, termed the apical ectoderm ridge (AER). Underlying this is a layer of undifferentiated mesenchymal cells termed the progress zone (PZ). Both are important in ensuring proximodistal (PD) outgrowth of the limb, via messenger proteins.

The human upper limb bud can be divided developmentally into four main zones: the stylopod, the zeugopod, the mesopod, and the autopod, which correspond approximately to the arm, forearm, wrist, and hand, respectively. Positional information regarding cell destination along the PD axis is conferred based on the length of time spent within the PZ. Cells exiting the PZ early are destined to form more proximal structures such as the humerus, while those with a longer residence develop into the more distal structures of the limb. Furthermore, each set of cells exiting the PZ differentiate and express different growth factors: the future stylopod expresses MEIS-1, the zeugopod cells express OXA11 and HOXD9, mesopod cells express HOXA1, and the autopod cells express HOXA13 and HOXD10–13.

32.3.1 Axes of Development

Limb growth and differentiation progress along three axes: PD, anteroposterior (AP), and DV. The PD axis refers to elongation of the limb, the AP axis defines radioulnar or preaxial and postaxial orientation, and the DV axis provides dorsal and palmar orientation. Development and differentiation of each axis is controlled by a separate signaling center. The AP axis is defined first, followed by the DV axis, and then the PD axis. The PD axis is controlled by the AER. It secretes WNT3 and fibroblast

growth factors (FGFs) including FGF-2, FGF-4, and FGF-8. This provides molecular control for the AER and for the differentiation and limb outgrowth through maintenance of FGF10 expression in the underlying mesoderm. This FGF10 expression sustains proliferation of the PZ mesodermal cells, which in turn are influenced by signaling centers to determine their fate. To maintain PD growth, combined expression of WNT and FGF is required.

AP axis development is controlled by the zone of polarizing activity (ZPA) in the posterior limb mesoderm. The ZPA expands limb width and promotes development of the posterior (ulnar) aspect of the limb through expression of SHH at the distal posterior border of the AER. The AER and ZPA form a reciprocal feedback loop to maintain SHH expression and therefore AP outgrowth.

DV axis growth is regulated by the secretion of WNT7A within the dorsal ectoderm. This induces the Lim homeodomain transcription factor LMX1B, which dorsalizes the underlying limb mesoderm. The ventral ectoderm secretes EN1 and LMX1. WNT7A has also been shown to maintain secretion of SHH (which influences many proteins) from the ZPA, thus linking outgrowth in all three axes.

Signaling centers coordinate control of downstream targets involved in development of the bony skeleton, vessels, muscles, and nerves of the upper limb. These can involve both common and asymmetric molecular pathways. For example, short-stature homeobox 2 (SHOX2) is upregulated in the proximal perichondrium to promote humeral elongation. SHOX, in comparison, is induced in the forearm perichondrium to regulate radioulnar elongation. Appropriate and systematic induction of downstream pathways through coordinated action at multiple signaling centers is essential in normal limb axis differentiation.

32.4 Skeletal Differentiation

At day 32, a hand plate is formed. All bones of the upper limb, except the distal part of the phalanges, form as mesenchymal condensations along the long axis of the limb during week 5. They undergo endochondral ossification in response to growth factors including FGFs, transforming growth factor beta (TGF- β), bone morphogenic proteins (BMP), Indian hedgehog (IHH), retinoids, and parathyroid hormone-related peptide (PTHrP). Chondrification occurs in a proximal to distal direction, and initially results in deposition of cartilage around the entire axis mesenchymal condensation, termed the perichondrium. A cartilaginous model of each bone is created, termed an anlage. Further chondrification is limited to the anlage of each endochondral bone. The mesenchyme in the interzones, which constitutes the sites of future joints, differentiates into fibrous connective tissue. Chondrification of each anlage is followed by ossification. This commences at the primary ossification center. Mesenchymal cells in the perichondrium differentiate into osteoblasts in response to TGF- β and secrete a mineralized bone matrix termed a primary bone collar around the circumference of the bone. This collar thickens as osteoblasts continue to differentiate in progressively more peripheral layers of the perichondrium. Osteoclastic activity facilitates bone remodeling. In the seventh week, the upper limb rotates so that the hands lie anteriorly and the elbows posteriorly.

Formation of the distal portion of the distal phalanges is different, by intramembranous ossification. This means they form directly from condensations of mesenchymal cells without the formation of cartilage intermediary.

Joints develop through repression of chondrogenesis at specific sites of future joints. At these zones, the wingless proteins WNT4 and WNT14 are expressed. The growth factor cartilage-derived morphogenetic protein 1 (CDMP1) is also expressed at these zones to repress chondrogenesis.

By birth, the limb diaphyses are completely ossified, but the epiphyses remain cartilaginous. Secondary ossification centers develop in these epiphyses and ossify. The persisting physis that lies between the epiphysis and metaphysis allows diaphyseal lengthening (growth).

32.5 Soft-Tissue Differentiation

Muscle fibers develop in the upper limb bud by the seventh week after fertilization, demonstrating migration and differentiation from superficial to deep and from proximal to distal. Innervation of the limb follows myocyte migration. SHH expression from the notochord induces motor neuron differentiation, while HOX transcription factors expressed in the spinal cord are thought to organize upper limb motor fibers into the lateral motor column and convey axon-targeting information to particular muscle groups.

The first axial vessel, the marginal vein, appears in the fifth week of development. Lymphatics develop to follow the veins. The arterial system, in contrast, is not seen until the 33rd day after fertilization. The brachial artery gives rise to the interosseous and median arteries. The median artery is the primary blood supply to the hand, and eventually gives rise to the ulnar and radial arteries. At this stage, the median artery usually regresses and supplies the median nerve alone.

The digital rays are initially syndactylized in utero. Hand plate formation is influenced by SHH and HOX transcription factors and BMP to create a posterior-to-anterior gradient. This allows interdigital signaling centers and digital anlagen to be established. The BMP gradient maps digit and interdigital fates on the hand plate and represses the AER. Digit separation occurs between the seventh and eighth weeks. Separation occurs by apoptosis at the interdigital mesenchyme under the influence of transcription factors including HOXD9, HOXD11, and HOXD13, progressing from distal to proximal.

At birth, despite a fully formed limb, myelination of the nerves is still incomplete.

32.6 Classification Systems

Congenital hand differences have traditionally been classified based on appearance. However, such descriptive terminology does not enlighten the surgeon regarding etiology, surgical approach and/or treatment, or overall prognosis. Dysmorphology can result from disruption at any time during the initial formative or later outgrowth stage of upper limb development.

32.6.1 Swanson Classification

The Swanson classification, first described in 1964, is based upon morphology. Categories include failure of formation,

failure of differentiation, duplication, overgrowth, undergrowth, constriction band syndrome, and generalized skeletal abnormalities. While its simplicity made it practical to use, as the understanding of the genetic and embryological basis of dysmorphology increased, a revised classification system taking these new developments into account was suggested.

32.6.2 Oberg–Manske–Tonkin Classification

The Oberg–Manske–Tonkin (OMT) classification (Box 32.1), first proposed in 2010, is based on the developmental biological and genetic basis of congenital limb anomalies rather than a purely descriptive framework. It separates malformations from deformations and dysplasias, and relates the diagnosis to the limb axis of development involved and the affected zone of the limb. Using such terminology allows each congenital condition to retain a surgical description and offers a framework for conveying the etiology of the anomaly while concomitantly remaining flexible to new developments and conditions. It also means that within the framework, if there are multiple anomalies (as found in syndromic presentations, e.g.), the condition may be classified under multiple diagnoses including both the syndrome and the upper limb anomalies. This is documented from most to least prominent, and allows documentation of all dysmorphic features, which may allow further identification of patterns of anomalies.

Box 32.1 Oberg–Manske–Tonkin Classification of Congenital Hand and Upper Limb Anomalies

Malformations

Failure of axis formation/differentiation: entire upper limb

1. Proximodistal axis
 - a) Brachymelia with brachydactyly
 - b) Symbrachydactyly
 1. Poland syndrome
 2. Whole limb excluding Poland syndrome
 - c) Transverse deficiency
 1. Amelia
 2. Clavicular/scapular
 3. Humeral
 4. Forearm
 5. Wrist (carpals absent/at level of proximal carpal row/at level of distal carpals) (with forearm/arm involvement)
 6. Metacarpal (with forearm/arm involvement)
 7. Phalangeal (proximal/middle/distal) (with forearm/arm involvement)
 - d) Intersegmental deficiency
 1. Proximal (humeral—rhizomelic)
 2. Distal (forearm—mesomelic)
 3. Total (phocomelia)
 - e) Whole limb duplication/triplication
2. Radioulnar (anteroposterior) axis
 - a) Radial longitudinal deficiency—thumb hypoplasia (with proximal thumb involvement)
 - b) Ulnar longitudinal deficiency
 - c) Ulnar dimelia
 - d) Radioulnar synostosis

- e) Congenital dislocation of the radial head
- f) Humeroradial synostosis—elbow ankyloses
3. Dorsoventral axis
 - a) Ventral dimelia
 1. Furhamm/Al-Awadi/Raas–Rothschild syndromes
 2. Nail–patella syndrome
 - b) Absent/hypoplastic extensor/flexor muscles
4. Unspecified axis
 - a) Shoulder
 1. Undescended (Sprengel)
 2. Abnormal shoulder muscles
 3. Not otherwise specified
 - b) Arthrogryposis

Abnormal axis formation/differentiation: hand plate

1. Proximodistal axis
 - a) Brachydactyly (no forearm/arm involvement)
 - b) Symbrachydactyly (no forearm/arm involvement)
 - c) Transverse deficiency (no forearm/arm involvement)
 1. Wrist (carpals absent/at level of proximal carpal row/at level of distal carpals)
 2. Metacarpal
 3. Phalangeal (proximal/middle/distal)
2. Radioulnar (anteroposterior) axis
 - a) Radial deficiency (thumb—no forearm/arm involvement)
 - b) Ulnar deficiency (no forearm/arm involvement)
3. iii Radial polydactyly
 - a) Triphalangeal thumb
 - b) Ulnar dimelia (mirror hand—no forearm/arm involvement)
 - c) Ulnar polydactyly
4. Dorsoventral axis
 - a) Dorsal dimelia (palmar nail)
 - b) Ventral dimelia (including hypoplastic/aplastic nail)
5. Unspecified axis
 - a) Soft tissue
 1. Syndactyly
 2. Camptodactyly
 3. Thumb in palm deformity
 4. Distal arthrogryposis
 - b) Skeletal deficiency
 1. Clinodactyly
 2. Kirner's deformity
 3. Synostosis/symphalangism (carpal/metacarpal/phalangeal)
 - c) Complex
 1. Complex syndactyly
 2. Synpolydactyly—central
 3. Cleft hand
 4. Apert hand
 5. Not otherwise specified

Deformations

Constriction ring sequence

Trigger digits

Not otherwise specified

Dysplasias**Hypertrophy**

1. Whole limb
 - a) Hemihypertrophy
 - b) Aberrant flexor/extensor/intrinsic muscles
2. Partial limb
 - a) Macrodactyly
 - b) Aberrant intrinsic muscles of hand

Tumorous conditions

1. Vascular
 - a) Hemangioma
 - b) Malformation
 - c) Others
2. Neurological
 - a) Neurofibromatosis
 - b) Others
3. Connective tissue
 - a) Juvenile aponeurotic fibroma
 - b) Infantile digital fibroma
 - c) Others
4. Skeletal
 - a) Osteochondromatosis
 - b) Enchondromatosis
 - c) Fibrous dysplasia
 - d) Epiphyseal abnormalities
 - e) Others

Syndromes**Specified**

1. Acrofacial dysostosis 1 (Nager type)
2. Apert
3. Al-Awadi/Raas-Rothschild/Schinzal phocomelia
4. Baller-Gerold
5. Bardet-Biedl carpenter
6. Catel-Manzke
7. Constriction band (amniotic band sequence)
8. Cornelia de Lange (types 1–5)
9. Crouzon
10. Down
11. Ectrodactyly-ectodermal dysplasia-clefting
12. Fanconi pancytopenia
13. Fuhrmann
14. Goltz
15. Gorlin
16. Greig cephalopolysyndactyly
17. Hajdu-Cheney
18. Hemifacial microsomia (Goldenhar syndrome)
19. Holt-Oram
20. Lacrimoauriculodentodigital (Levy-Hollister)
21. Larsen
22. Leri-Weill dyschondrosteosis
23. Moebius sequence
24. Multiple synostoses
25. Nail-patella
26. Noonan

27. Oculodentodigital dysplasia
28. Orofacial digital
29. Otopalatal digital
30. Pallister-Hall
31. Pfeiffer
32. Poland
33. Proteus
34. Roberts-SC phocomelia
35. Rothmund-Thomson
36. Rubinstein-Taybi
37. Saethre-Chotzen
38. Thrombocytopenia absent radius
39. Townes-Brock
40. Trichorhinophalangeal (types 1–3)
41. Ulnar-mammary
42. VACTERL syndrome association

Others

Abbreviation: VACTERL syndrome, vertebral anomalies, anal atresia, cardiac abnormalities, tracheoesophageal fistula, renal agenesis, and limb defects.

Note: The specified syndromes detailed in the table are those considered most relevant based on the latest OMT revised classification and IFSSH publications. However, many other syndromes have a limb dysmorphology component and are categorized under “B. Others.”

As already alluded to, the OMT classification uses dysmorphology terminology to incorporate etiology and developmental biology concepts into the framework of classification. These terms are malformation, deformation, and dysplasia. A malformation is an abnormal formation of a body part. Deformation refers to an insult occurring after normal formation. A dysplasia is an abnormality in size, shape, and organization of cells within a tissue. Separating conditions based on whether they result from a malformation, deformation or dysplasia provide a solid basis for etiological classification. Malformations are further subdivided according to the predominantly affected axis of development, and the zone of the limb involved. The use of dysmorphology terminology allows this framework the capacity to be adapted and refined as our understanding of limb development increases, as demonstrated by its revisions and extensions since initial publication.

In 2014, the International Federation of Societies for Surgery of the Hand (IFSSH) Scientific Committee on Congenital Conditions recommended adoption of the OMT classification over the Swanson classification. This is with the reservation that the classification would be reviewed, once in common use, to reassess its utility. Indeed, it has already been extended with several modifications since its inception. It remains to be seen as to its applicability in everyday clinic situations or whether its usefulness is more widespread within the research forum. Currently, neither the Swanson nor the OMT classification determines or suggests surgical management, but if future developments include the use of pre-implantation gene modification techniques, this may change.

32.7 Disruptions to Axis Formation/Differentiation

Understanding of how developmental disruption and interruption of signaling pathways lead to specific hand differences is continually improving, so just a few examples are included here since greater detail is to be incorporated within subsequent chapters.

Intersegmental phocomelias have been associated with SHOX and SHOX2 with abnormalities in PD outgrowth. AP deficiencies have been noted with reduced FGF function, resulting in a range of phenotypes including Apert syndrome. Progressive loss of SHH expression or targeted temporal loss of SHH signaling during development generates a spectrum of ulnar longitudinal deficiencies. When associated with reduced FGF expression through an interrupted SHH-FGF loop, this may result in radial deficiencies, particularly involving the thumb, which explains why most ulnar longitudinal deficiencies are associated with both preaxial and postaxial deficiencies within the hand plate. Brachydactyly type B1 has been associated with a mutated receptor tyrosine kinase (ROR2) and WNT signaling.

Reduction or loss of WNT7A or LMX1B expression in the dorsal mesoderm results in impaired dorsalization, with abnormal elbow and nail formation such as seen in nail-patella syndrome.

32.7.1 Soft-Tissue Deficiencies

Ectopic overexpression of BMP inhibitors in the limb mesenchyme and persistent or overexpressed FGF in the AER can result in inhibited interdigital apoptosis with resultant syndactyly. Mutations in BMP inhibitors are associated with syndactyly, polydactyly, and joint synostoses. GLI3 and HOXD13 gene mutations can cause these anomalies, but the pattern depends on which specific signals are interrupted so that identical genotypes may produce quite significantly different phenotypical appearances and hence the need for a classification based on etiology and pathogenesis rather than morphology.

32.8 Conclusion

The embryogenesis of the upper limb relies on a relatively complex and closely regulated sequence of transcription factors and

growth factors to ensure normal development. Limb embryogenesis is best thought of in terms of three axes of development: PD, AP, and DV. Appreciation of normal embryology allows a better understanding of the underlying etiology of congenital limb anomalies, and therefore can aid an appreciation of the future management, including operative approaches, and prognosis. The OMT classification takes these aspects into consideration and offers a logical framework for this wide spectrum of anomalies. While conditions may require classification under multiple diagnoses including both the syndrome and the upper limb anomalies, this allows comprehensive documentation of all anomalies. As the genetic basis of upper limb embryology is further elucidated, review and further streamlining of this framework may be achieved in the future.

32.9 Key Points

The upper limb develops between the fourth and eighth weeks after fertilization from the x and y.

- Development should be considered in terms of the DV, AP, and PD axes.
- The OMT classification of congenital hand and upper limb anomalies incorporates our understanding of etiology, molecular genetics, and developmental biology to help organize the main groups and subgroups of congenital abnormalities and syndromes.
- Classification can be based on the broad themes of malformations, deformations, and dysplasias, with cross-referencing for abnormalities with an associated syndromic component.

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33 Disorders of Failure of Formation in the Upper Limb

Dariusz Nikkhah and Bran Sivakumar

Summary

Disorders of failure of formation of the upper limb include: transverse arrest, radial and ulnar longitudinal deficiency, and central ray deficiency. The conditions are usually sporadic, but may be associated with several syndromes. Management is based on the type and severity of the deformity.

Keywords: longitudinal deficiency, transverse arrest, central ray deficiency, cleft hand

33.1 Transverse Deficiencies

Congenital transverse arrest is a rare abnormality that commonly occurs at the level of the wrist or proximal forearm. Transverse arrest is usually sporadic without a genetic inheritance; its incidence has been reported as 6% of all congenital upper limb deficiencies.

33.1.1 Diagnosis of Transverse Arrest

Transverse arrest can occur at multiple levels, from the humeral down to the metacarpal level of the upper limb. Transverse arrest above the elbow is rare. The distal stump is usually rounded and often contains some nubbinlike hand/digital remnants (► Fig. 33.1). Areas of dimpling are sometimes present where the skin is tethered to the end of hypoplastic skeletal structures. The condition is most often unilateral and more often affects the left side. It has a 3:2 female predominance.

33.1.2 Treatment of Transverse Arrest

Regular detailed assessments of function and psychosocial well-being are required prior to embarking on any treatment given these children often adapt remarkably well to their limb differences.



Fig. 33.1 Transverse arrest at proximal forearm level. Rudimentary hand remnants visible at tip of rounded stump.

The management of transverse arrest varies according to the level of involvement. In above-elbow transverse deficiencies, the main treatment options involve prostheses. However, in unilateral cases, compliance to prostheses can be poor in part due to a lack of sensory feedback from the devices. However, a variety of prostheses are available for trial and range from static to dynamic, some of which can be controlled by remaining skeletal structures or myoelectric impulses.

Surgical options for proximal transverse deficiencies include the removal of excess functionless parts such as digital nubbins or stump revision to facilitate prosthetic fitting. At the forearm level, few surgical options exist; one historic option described is the Krukenberg procedure. The radius and ulna are separated to create a basic prehensile pattern between the two bones. This procedure is not recommended because the cosmetic appearance is very poor and the separation of the forearm bones can prevent the effective use of prostheses.

Vascularized second-toe transfer is an option in children with carpal and metacarpal hands. In children with carpal hands, second toes can be transferred to the positions of the first and fifth digits. In the absence of digits, the transfer of two second toes in this way allows the creation of a basic prehension pattern. This tends to be more effective in cases where the child is able to cup their palm.

Distal cases of transverse arrest are often referred to as symbrachydactyly. Treatment options in these cases include web deepening, metacarpal transposition, distraction lengthening of existing skeletal parts, vascularized second-toe transfers, and nonvascularized toe phalangeal transfers. Preservation of the phalangeal periosteum and minimization of tension within the recipient soft-tissue envelopes in the hypoplastic digits are key to long-term survival of nonvascularized toe phalangeal transfers.

33.1.3 Outcomes and Complications

Careful assessment of cases is vital to prevent unnecessary intervention given these children adapt remarkably well. All viable treatment options must be explained to the child and family. The unpredictability of prosthesis use in unilateral cases must be emphasized. In more distal cases, at the carpal and metacarpal level, the creation or upgrade of a prehension pattern can significantly improve hand functionality and increase a child's bimanual repertoire.

Donor site issues appear to be common in cases of nonvascularized toe phalangeal transfers particularly when children reach their adolescent growth spurt. However, techniques to minimize these complications such as the use of replacement iliac crest bone grafts have been shown to be effective.

Distal transverse deficiency is often an ideal indication for free vascularized toe transfer. Excellent outcomes have been seen in terms of function, cosmesis, and psychosocial well-being with little donor site morbidity in large series with significant long-term follow-up.

33.2 Longitudinal Deficiencies

33.2.1 Radial Longitudinal Deficiency

Radial Longitudinal Deficiency (RLD) represents a spectrum of anomalies, which can involve the entire preaxial side of the upper limb. In the past, a variety of terms have been used to refer to the condition, including radial club hand, radial hypoplasia, and meromelia. RLD is the most common out of the longitudinal deficiencies and occurs in 1/30,000 cases. Unilateral cases are more common with a R:L predilection of 2:1. Genetic and environmental factors such as thalidomide have been shown to play a role in RLD.

RLD can be linked with other anomalies and syndromes, and therefore screening is essential (Box 33.1). Bilateral cases of RLD are more likely to have a syndromic association.

Box 33.1 Cardiac, Renal, and Hematological Syndromes Associated with Radial Longitudinal Deficiency

- Holt–Oram syndrome: vertebral anomalies, anal atresia, cardiac abnormalities, tracheoesophageal fistula, renal agenesis, and limb defects
- VACTERL syndrome: vertebral anomalies, anal atresia, cardiac abnormalities, tracheoesophageal fistula, renal agenesis, and limb defects
- Fanconi syndrome: autosomal recessive condition with aplastic anemia
- TAR: autosomal recessive condition with thrombocytopenia and absent radius

33.2.2 Diagnosis of Radial Longitudinal Deficiency

As a result of skeletal deficiency and soft-tissue abnormalities in RLD, the hand adopts a radially deviated and abnormally flexed posture at the end of a hypoplastic forearm (► Fig. 33.2).



Fig. 33.2 Image of patient with radial longitudinal deficiency, demonstrating short limb with stiff elbow and radial deviation and flexed posture of the wrist joint. Note absence of the thumb and widening of the second webspace.

Complete assessment of the upper limb is important given that abnormalities can occur along its entire length from the shoulder to the radial side of the hand. The upper arm may be shorter with distal humeral epiphyseal involvement and in some cases a stiff elbow. The forearm is shorter to varying degrees and the severity of skeletal hypoplasia is classified into four grades based on radiographic assessment according to the system described by Bayne and Klug (► Table 33.1). Type 1 represents a short radius and type 2 a hypoplastic radius with involvement of the proximal and distal epiphyses. In type 3, the distal part of the radius is replaced by a tethering fibrous anlage and in type 4 there is complete radial aplasia. The main factors contributing to radial deviation are hypoplasia and mal-insertion of a combined muscle mass of FCR, ECRB, ECRL, and BR, radial displacement of the median nerve, tightened deep fascia, and in some type 3 cases a radial anlage. The forearm–wrist articulation is often unstable and nonfunctional with hypoplasia of the radial side of the carpus. Within the hand, the thumb is hypoplastic or most often absent. The index and middle fingers are also affected with varying degrees of hypoplasia and stiffness.

33.2.3 Associated Syndromes

There are a number of syndromes linked to RLD; these include VACTERL (vertebral anomalies, anal atresia, cardiac abnormalities, tracheoesophageal fistula, renal agenesis, and limb defects) association, TAR (thrombocytopenia and absent radius) syndrome, and Holt–Oram and Fanconi anemia (Box 33.1). These syndromes, which occur in approximately one-third of patients with RLD, confer a poorer prognosis and require early detection and treatment. Complete assessment of the limbs, cardiac, renal, spinal, and hematological systems must be carried out.

33.2.4 Treatment of Radial Longitudinal Deficiency

The main aims of treatment in RLD are to create stable alignment of the hand and carpus on the end of the ulna, maximize function, and optimize growth potential.

Treatment begins with a regime of regular stretching and splinting under the guidance of a hand therapist. Physiotherapy aims to address the shortened deviating soft-tissue structures on the radial side of the wrist and at the same time maintain digit mobility and strength.

Subsequent surgical intervention depends on the severity of the hypoplasia and the child's overall functionality. In the

Table 33.1 Bayne classification of radial longitudinal deficiency

GR	Radial longitudinal deficiency
I	Short distal radius
II	Hypoplastic radius
III	Partial absence of radius
IV	Total absence of radius

Source: Bayne and Klug (1987).

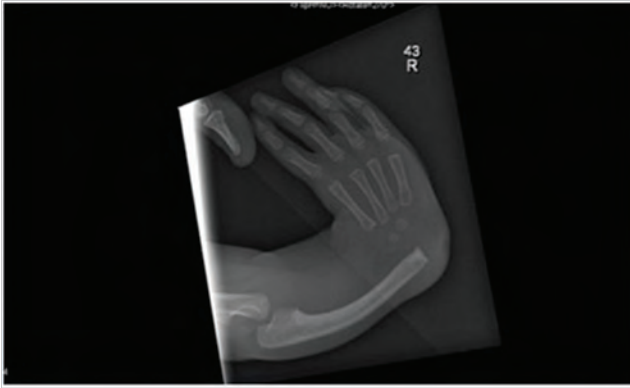


Fig. 33.3 Uniplanar soft-tissue distraction device in situ; it is fitted onto the radial aspect of the limb.

majority of type 1 cases where the wrist is stable and easily corrected to neutral, no intervention is required. However, if a tendency for radial deviation remains, a tendon transfer of the tight combined dorsoradial muscle mass to the ulnar side of the carpus may be required. Type 2 hypoplastic RLD cases are more challenging to treat. Distraction lengthening of a small radius can be effective but often needs repeating to keep up with growth. Type 3 and 4 cases are best dealt with by soft-tissue distraction of the short radial deforming structures followed by a wrist centralization/radicalization procedure to create a stable ulna–carpal equilibrium.

33.2.5 Soft-Tissue Distraction

Kessler first described the use of gradual distraction lengthening of soft tissues in RLD. This technique facilitates definitive wrist correction and minimizes the need for skeletal shortening. Since Kessler's first description, a range of different distraction devices have been successfully applied to cases of RLD including uniplanar and ring fixator systems. Our preference is a uniplanar device with a central coaxial hinge placed on the radial side of the limb and secured by pin fixation into the ulnar proximally and into the radial metacarpals distally (► Fig. 33.3). Distraction is performed at approximately 0.5 to 1 mm a day for 6 to 8 weeks and stopped once the third metacarpal base is aligned with the distal ulna in preparation for the wrist corrective procedure.

33.2.6 Wrist Correction in Radial Longitudinal Deficiency

Wrist correction is achieved through either a centralization or a radicalization procedure. Centralization involves fixation of the distal ulna in line with the third metacarpal in a central slot created in the proximal carpal row. As a result of the fusion to the proximal row, centralization results in more stability but less wrist motion and shorter limb when compared to radicalization. Radicalization involves stabilization of the carpus of the ulna in line with the second metacarpal without a carpal slot. Both procedures require transfer of the dorsoradial muscle mass into the base of the fifth metacarpal to convert a deforming radial force into corrective stabilizing dorso–ulnar one. A

radicalization is more dependent on the quality of this tendon transfer for long-term stability. Both types of correction are supported with longitudinal K-wire fixation for a period of 6 to 12 months.

33.2.7 Microvascular Reconstruction in Radial Longitudinal Deficiency

An alternative to centralization/radicalization in RLD is reconstruction of a stabilizing radial buttress using vascularized fibula or metatarsophalangeal transfers. This technique is gaining popularity in a number of units around the world, and long-term follow-up of this approach is ongoing.

33.2.8 Outcomes and Complications of Radial Longitudinal Deficiency

Soft-tissue distraction has revolutionized treatment of RLD. It has allowed the optimization of limb length, growth potential, and wrist mobility (ulna–carpal) in type 3 and 4 cases.

A radicalization, although it preserves motion and length, is inherently more unstable than a centralization and as a result is more vulnerable to long-term recurrent radial deviation. Therefore, we advocate a centralization in cases where the quality of the dorsoradial muscle mass or ulna–carpal joint congruity does not lend itself to the stable equilibrium required for a radicalization.

A formal wrist arthrodesis is a salvage procedure for children nearing skeletal maturity in cases of recurrent deviation and resultant poor function despite treatment.

33.3 Central Ray Deficiency

Central longitudinal failure of formation (congenital limb defect [CLD]) also known as cleft hand represents a spectrum of deformities that involve hypoplasia or absence of the central rays of the hand. Children with this condition typically function well and have been described by Flatt as “a functional triumph and social disaster.” Typical presentation includes the presence of a “V”-shaped central cleft, malpositioning of adjacent rays with narrowing of the first webspace (► Fig. 33.4). It is distinct from symbrachydactyly, which typically presents with a broad “U”-shaped cleft and bordering digital hypoplasia. CLD is an autosomal dominant condition with variable expressivity. CLD can occur in isolation, but the majority of patients present with cleft hands and feet, a spectrum termed split hand (SH) and split foot (SF) complex.

33.3.1 Diagnosis of Congenital Limb Defect

A wide range of cleft hand morphology exists ranging from simple soft-tissue clefts without digital absence, through varying degrees of central ray hypoplasia to complete suppression of all bony elements of the hand except for the little finger. A number of different classification systems exist to describe the range of deficiencies seen in CLD, the most useful of which was put forward by Manske in 1995. This system is based on the quality of

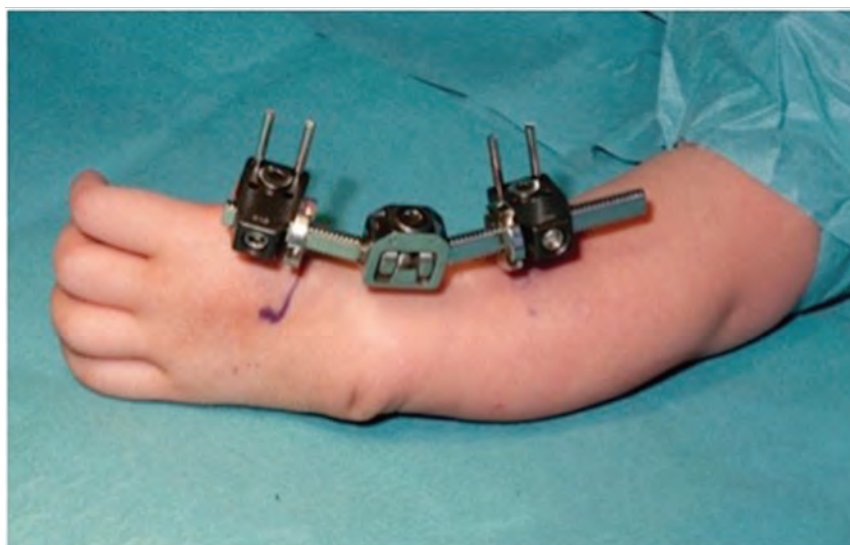


Fig. 33.4 Image of patient with central longitudinal deficiency. Note absence of distinct central ray, merging of long and ring finger with malposition of the index finger and narrowing of the first webspace.

Table 33.2 Manske first webspace classification of cleft hand

GR	Manske first webspace classification
I	Normal web: no treatment
IIB	Mildly narrowed web: four-flap Z-plasty
IIA	Severe narrowing: palmar- or dorsal-based flap
III	Syndactylized thumb: palmar or dorsal rotation flap
IV	Merged webspace: cleft deepening and rotational osteotomies
V	Absent web: microsurgical toe transfer

the first webspace and radial digits, and provides a better indication of likely function than classifications that examine the central ray deficiency itself (► Table 33.2). The classification ranges from grade 1 where there is no significant narrowing of the first web, through worsening grades of first web narrowing, syndactyly, and merging (grades 2–4) to complete absence of the thumb and radial digits (grade 5). The classification system also serves as a useful guide for treatment (► Fig. 33.5).

33.4 Treatment of Cleft Hand

Close monitoring of patients with CLD is important given that many children adapt well to a central deficiency and show no significant functional impairment. However, some cleft hands do require some functional optimization; for example, if the index finger sits too close to the thumb, it may be bypassed in pinch in preference for the ring finger. Transposition of the index finger into the long finger position allows creation of a wider first webspace and greater inclusion of the index finger into grasp and pinch. Other major indications for operative intervention include narrowing/syndactyly of the first webspace and severe flexion contracture of the digits. Timing of surgery varies according to functional need. Early intervention is advocated in cases of first web syndactyly to prevent thumb–index length discrepancy from affecting growth.



Fig. 33.5 Type 4 radial longitudinal deficiency with complete absence of the radius and bowing of the ulna with no functional wrist joint. The hand and carpus are radially deviated and there is complete absence of the thumb.

33.4.1 Surgical Techniques

A simple soft-tissue cleft can be closed with a small commissural flap, which allows the creation of a natural “U” shape within the webspace. Within deeper clefts, a range of different techniques utilizing local random pattern flaps of volar or dorsal skin have been employed to resurface the newly widened first webspace following transposition of a malpositioned index finger. Problems of distal flap necrosis and subsequent web



Fig. 33.6 Surgical correction of typical cleft hand using Upton technique. Note the transposition of the index finger into the long finger position with widening of the first webspace. A clinodactyly correction in the ring finger has also been carried out.

contracture have prompted evolution of these techniques away from long random pattern flaps. Our preferred method is that described by Upton in 2010 in which the volar and dorsal skin are peeled back via a racket incision around the index finger with straight-line extensions (between the glabrous and dorsal skin) along the cleft and first webspace. The index finger is then transposed, problematic aberrant anatomy addressed, and the skin redraped to create a new broad first webspace and a natural second webspace contour with the help of a commissural flap from the radial side of the ring finger (► Fig. 33.6). The new position for the index finger is held using K wires into the base of the middle finger metacarpal remnant or distal carpal row. Preservation of the adductor pollicis origin, if present, is important in order to optimize key pinch. Reconstruction of the intermetacarpal ligament can be achieved through simple sutures alone, circumferential sutures around adjacent metacarpals, or through the union of adjacent A1 pulleys.

Adjacent severe flexion contractures of other digits can be addressed along the principles outlined for camptodactyly treatment.

33.4.2 Outcomes and Complications of Cleft Hand Correction

Surgery must be carefully considered in CLD because function should always take priority over aesthetics. Complications of flap contracture within the webspaces have been reduced through avoidance of long random pattern flaps. Long-term follow-up in these cases is essential as issues can arise as a result of differential growth between affected and normal areas, for example, between long index fingers and short and sometimes contracted adjacent digits. Problems of progressive joint contractures must be addressed early.

33.5 Ulnar Longitudinal Deficiency

Ulnar longitudinal deficiency (ULD), referred to in the past as ulnar club hand, is the rarest of the longitudinal deficiencies (1–

7.4 per 100,000 births). As with RLD, it can affect the entire length of the upper limb. However, unlike in RLD, the wrist is usually more stable and the elbow more severely affected in ulnar deficiencies. ULD is frequently associated with other musculoskeletal abnormalities and sometimes linked to syndromes affecting the hematopoietic, gastrointestinal, genitourinary, and cardiopulmonary systems.

33.5.1 Diagnosis of Ulnar Longitudinal Deficiency

ULD is graded into four main types according to the Bayne classification. Grade 1 refers to a hypoplasia of the ulna. Grade 2 is the most common and represents a partial aplasia of the ulna with bowing of the radius and sometimes a dislocation of the radial head. In grade 3, the ulna is entirely absent and there is again significant bowing of the radius with a radial head dislocation. Finally, in type 4, there is a radiohumeral synostosis alongside complete aplasia of the ulna. More recently, patients with an isolated ulnar-sided deficiency of the hand and digits in the presence of a normal forearm have been referred to as having a type 0 ULD. Patients tend to present with varying degrees of ulna hypoplasia and forearm bowing (► Fig. 33.7). In some patients, this results in a “hand behind the back” posture, which is particularly evident in grade 4 cases. The wrist deformities seen in ULD are secondary to stunted growth of the ulnar side of the distal radial epiphysis and in some cases a tethering effect of an ulna anlage. In the majority of cases, it is the hand deformities that impact on patient function the most. The severity of these hand abnormalities appears unrelated to the degree of the forearm, elbow, and upper arm deficiency. Furthermore, in over 50% of cases of ULD, there is an abnormality affecting the radial side of the hand, the functional significance of which prompted the creation of a supplementary classification by Manske examining the state of the thumb and first webspace in ulnar deficiencies (► Table 33.3).

33.6 Treatment in Ulnar Longitudinal Deficiency

Conservative treatment involving splinting and stretching in the first year of life can be helpful in some cases of ULD.

The majority of surgery in cases of ULD are focused on the hand because this appears to hold the greatest functional benefit. Commonly performed procedures include syndactyly releases, rotational osteotomies to facilitate prehension, webspace deepening in particular of the first webspace, thumb reconstructions, and pollicizations.

With regard to the forearm in Bayne grades 2 and 3, there is often a functional benefit from excision of the fibrous ulnar anlage to prevent progressive deformity of the radius. If there is significant radial deformity, the excision of the anlage can be performed alongside a corrective osteotomy of the radius. In some grade 3 and 4 cases, creation of a one-bone forearm from the distal radius and proximal ulna allows for simultaneous stabilization of the wrist and elbow, removal of an obstructive dislocated radial head, and optimization of forearm length. It is best performed in patients with radial head dislocation, forearm instability, and marked limitation of elbow extension.



Fig. 33.7 Image of ulnar longitudinal deficiency with typical shortened forearm appearance with deficiency of ulnar digits. Note previous first web widening scarring.

Table 33.3

GR	Manske classification of ulnar deficiency, 1997
I	Normal first web and thumb
II	Mild first web and thumb deficiency
III	Moderate to severe first web and thumb deficiency
IV	Absent thumb

Surgery to the elbow is sometimes required in cases of grade 4 ULD where there is a radiohumeral synostosis and the limb adopts a “behind the back” posture. In these cases, a de-rotational osteotomy through the synostosis allows for a more functional placement of the forearm and hand in front of the body.

33.6.1 Outcomes and Complications of Ulnar Longitudinal Deficiency Surgery

There are few long-term studies looking at subjective and objective outcomes in ULD most probably due to the rarity of the condition. Humeral and radial osteotomy surgery is useful to treat those children with poor spatial positioning of the limb. However, in the majority of ULD patients, the greatest functional gains are from surgery to the hand.

Minor complications include web creep after syndactyly release and more major complications include delayed union, malunion or nonunion, or failure to correct positional deformities. Iatrogenic injury to the posterior interosseous nerve is a risk in the creation of a one-bone forearm (Sénès and Catena 2012).

33.7 Key Points

- Transverse arrest often does not require surgical intervention as children adapt well.
- RLD is associated with many coexisting syndromes and patients should be screened for cardiac, hematological, and renal abnormalities.
- Soft-tissue distraction has revolutionized the management of RLD, reducing recurrence, optimizing growth potential, and facilitating surgical wrist correction.
- Classically function is good within cleft hands, but aesthetics can be poor.
- The majority of the surgery in ulnar longitudinal deficiency is directed at the hand.

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34 Thumb Hypoplasia

Amir H. Taghinia and Joseph Upton

Summary

Thumb hypoplasia can cause significant loss of function because it is the most important digit of the hand. Most patients require surgical intervention which is based on the type of hypoplasia. Treatment ranges from first webspace widening to index finger pollicization.

Keywords: thumb, hypoplasia, classification, pollicization

34.1 Introduction

The thumb is the prime ray of the hand. Its size, position, mobility, and relationship to other digits is critical for function. In the first few months of life, the thumb is adducted and flexed within the palm. Large object grasp starts around 6 months of age, and small object pinch starts around 9 months of age. By this time, the first ray has gained independence from the palm and by a year of age, it has become the most important ray of the hand. In the young toddler, the thumb is used creatively and independently to explore and manipulate the environment.

The thumb is considered hypoplastic if any portions of the ray including bony or soft-tissue components are underdeveloped or absent. Children with thumb hypoplasia can suffer from other associated conditions that affect the heart, blood, skeleton, and other systems. The widely accepted classification system for thumb hypoplasia was initially proposed by Blauth and later modified by Manske. This system is concise and practical as it guides surgical treatment (► Fig. 34.1).

With the exception of mild hypoplasia, most patients with thumb hypoplasia require surgical intervention. Treatment ranges from simple procedures such as first webspace widening to more complex procedures such as index finger pollicization. Over the past 30 years, there have been refinements in surgical techniques that improve both appearance and function. Recent

advances in outcome instruments hold promise for better functional delineation of the impact of surgical interventions for these conditions.

34.2 Diagnosis

Correct diagnosis starts with a thorough physical examination and plain radiographs. Although it is difficult to accurately assess ligament stability and active function in a newborn, an accurate assessment of deficient anatomy is possible. Accordingly, a preliminary diagnosis can be given early on and then modified as the child grows. Once the child starts to use the hand, a correct diagnosis is much easier to establish. Physical examination and radiographs should guide classification of the thumb using the Blauth system (► Fig. 34.1).

34.2.1 Associated Conditions

It is critical for the hand surgeon to be aware of conditions that are associated with thumb hypoplasia. Oftentimes—especially in those with blood conditions—these children manifest radial deficiency as their only physical difference. Accordingly, the hand surgeon can be instrumental in diagnosing associated conditions. These conditions include, but are not limited to, Fanconi anemia, Holt–Oram syndrome, and VACTERL (vertebral anomalies, anal atresia, cardiac abnormalities, tracheoesophageal fistula, renal agenesis, and limb defects) complex.

Type I: Mild Hypoplasia

In this mildest type of hypoplasia, the thumb is thinner and shorter than normal. The joints show normal range of motion, and flexion and extension creases are easily visible. There may be slightly smaller thenar intrinsic muscles, mainly the abductor pollicis and opponens pollicis; however, all of the

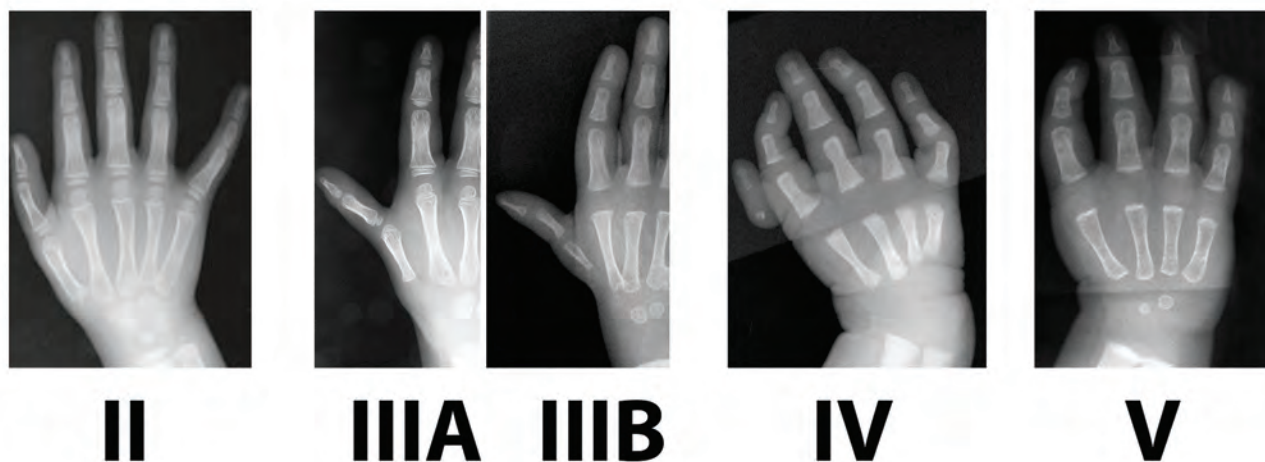


Fig. 34.1 Radiographical classification of thumb hypoplasia. Type I hypoplasia, the mildest severity, is not shown. The main distinguishing feature between type IIIA and IIIB hypoplasia is the presence of a functional carpometacarpal joint.

intrinsic muscles are present. The joints are stable. The first webspace deficiency may be slight to moderate.

Type II: Moderate Hypoplasia

In this type of hypoplasia (► Fig. 34.2), the bones of the ray are present but small. The radial carpal bones (trapezium and scaphoid) may be small and cause slight radial deviation of the hand. The first webspace is narrow and the thumb adducted. The metacarpophalangeal (MP) joint collateral ligaments are lax and the median-innervated thenar muscles are hypoplastic or absent. There may be extrinsic flexor and/or extensor deficiencies and these usually manifest by the lack of flexion or extension creases.

Type III: Severe Hypoplasia

The degree of thumb skeletal shortening and joint laxity is much more severe in these hands. These thumbs have been divided into subtypes A and B, based on the length of the

metacarpal and stability of the carpometacarpal (CMC) joint. In type A (► Fig. 34.3), the CMC joint is stable, the metacarpal is narrow, and the child uses the thumb. In type B (► Fig. 34.4), the CMC joint is unstable, the metacarpal is incomplete, and the child does not use the thumb. Type IIIA thumbs have severely hypoplastic thenar muscles and usually lack an extrinsic flexor. The collateral ligaments at the MP joint are lax and prevent sustained large object grasp—children usually use the head of the metacarpal for grasp. The extrinsic muscles insert abnormally and conspire to act primarily as radial deviators and not as primary flexors or extensors. This is known as “pollex abductus.” Many anatomical variations of this anomaly exist, but the functional result is identical: when the extrinsic muscles activate, the thumb radially deviates. Type IIIB thumbs share many of the same characteristics as type IIIA thumbs with greater degree of hypoplasia. In addition, the CMC joint is unstable in these thumbs. Most children with these thumbs will not use the thumb routinely; instead, they automatically use the index and middle fingers.

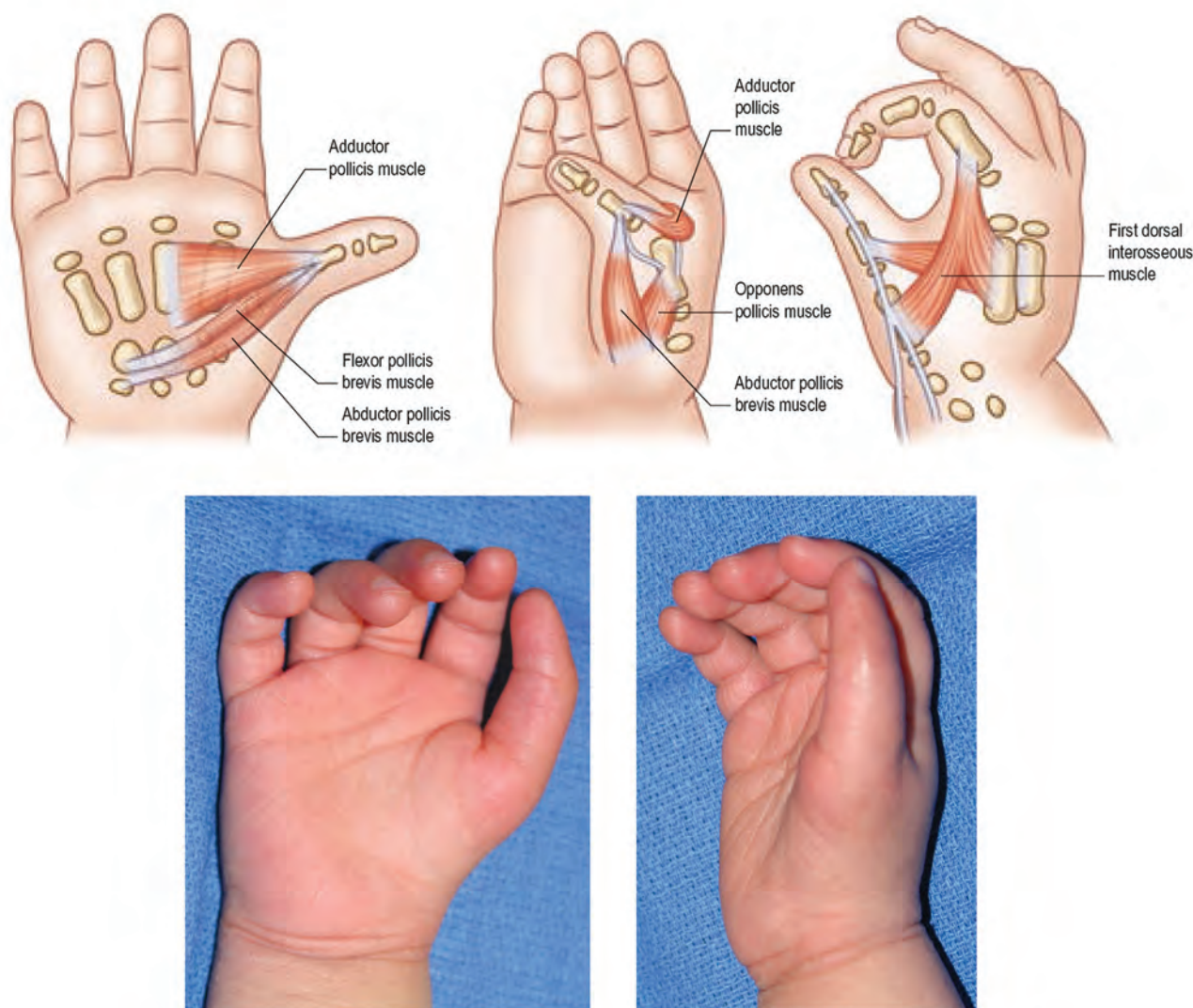


Fig. 34.2 Type II thumb hypoplasia.

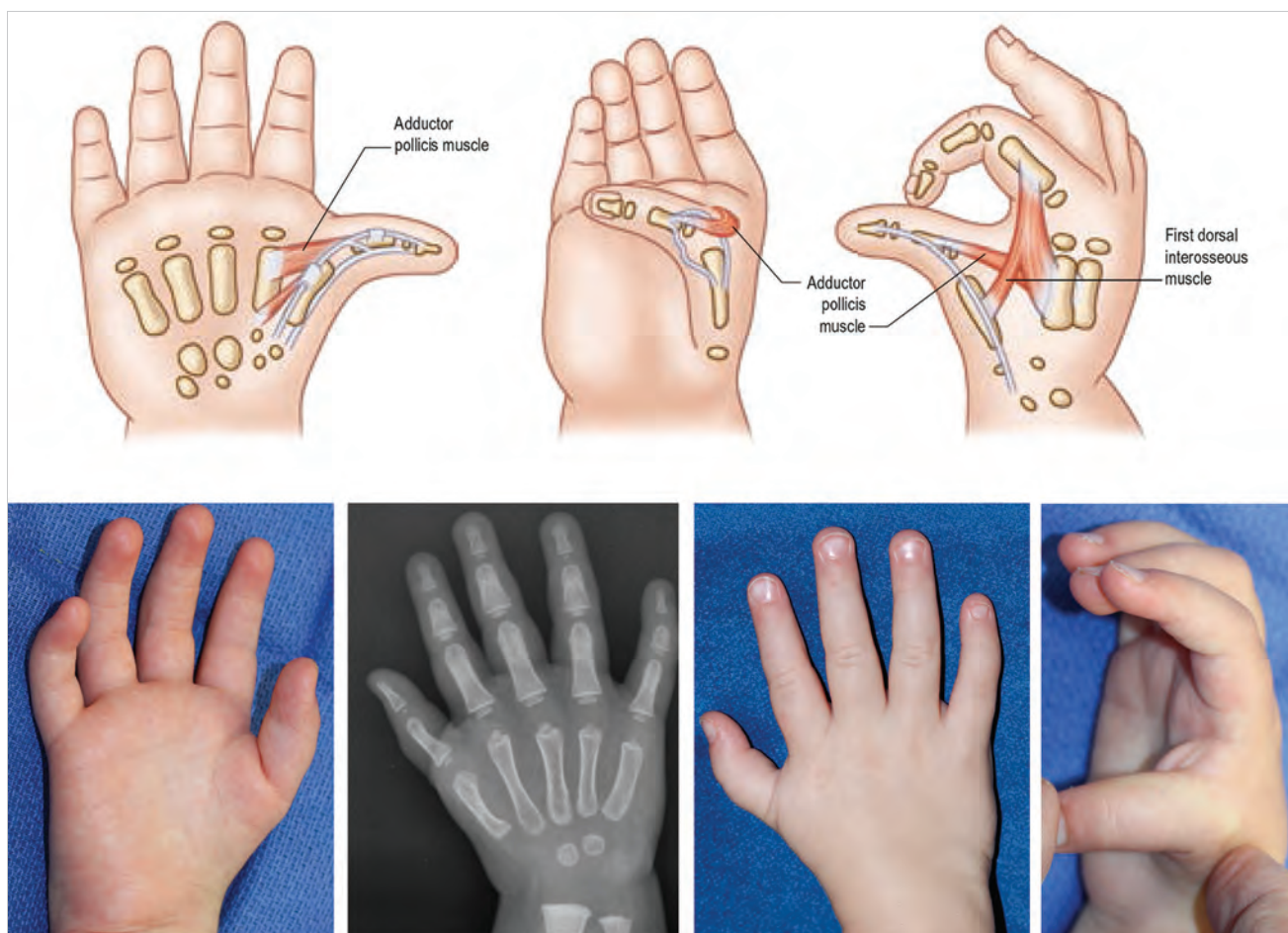


Fig. 34.3 Type IIIA thumb hypoplasia.

Type IV: Floating Thumb

The floating thumb (pouce flottant, French; pendeldaumen, German) is only attached to the hand by a soft-tissue pedicle that contains a neurovascular bundle (► Fig. 34.5). There is no metacarpal. The thumb has a rudimentary nail and one or two small phalanges. It is functionless.

Type V: Aplasia

In this type of hypoplasia, the thumb is completely absent (► Fig. 34.6). The radial side of the hand is hypoplastic. There is often an associated deficiency of the radius; and if so, there may be concomitant hypoplasia or stiffness of the index finger. If the index finger is normal, many patients will autopronate the index finger and scissor-grasp with the middle finger. The pulp of the index finger widens and the digit pronates, sitting in a more abducted position and thus widening the intermetacarpal space.

Other Types: Five-Fingered Hand

In this type, the most radial digit looks and functions like a finger (► Fig. 34.7). It usually lies in the same plane as the ulnar four digits and is nonopposable. Severe deficiency of the first

webpace is present. The metacarpal has a distal growth center and the intrinsic muscles are those that typically motor a finger, not a thumb.

Other Types: Radial Polydactyly

Many have observed that each partner of a radial polydactyly thumb is hypoplastic to varying degrees. The ulnar partner is less affected. In these thumbs, opponensplasty is usually not needed because the disposed radial partner usually presents a strong intrinsic muscle. The webspace may need widening in more proximal arborizations. Extra phalanges and pollex abductus abnormalities should be recognized.

34.3 Nonoperative Treatment

Conservative treatment alone is advocated for the most minor types of thumb hypoplasia. Splinting and stretching exercises are most useful early on in infancy when the soft tissue is pliable. In those patients who bypass a less functional thumb, one can try to encourage use of the thumb by buddy taping the index and middle fingers. This technique is useful early on in patients who have had thumb reconstruction or even pollicization.

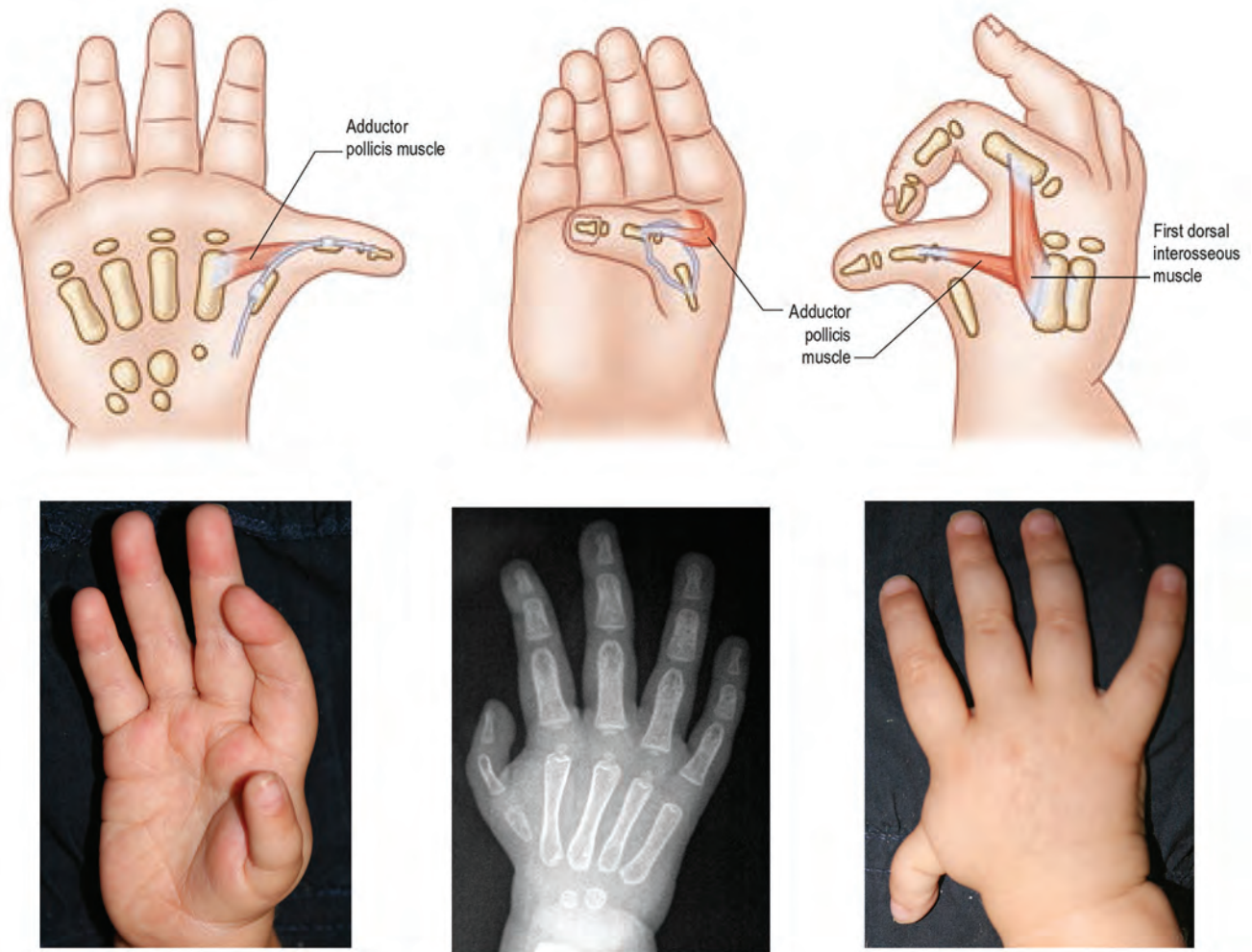


Fig. 34.4 Type IIIB thumb hypoplasia.

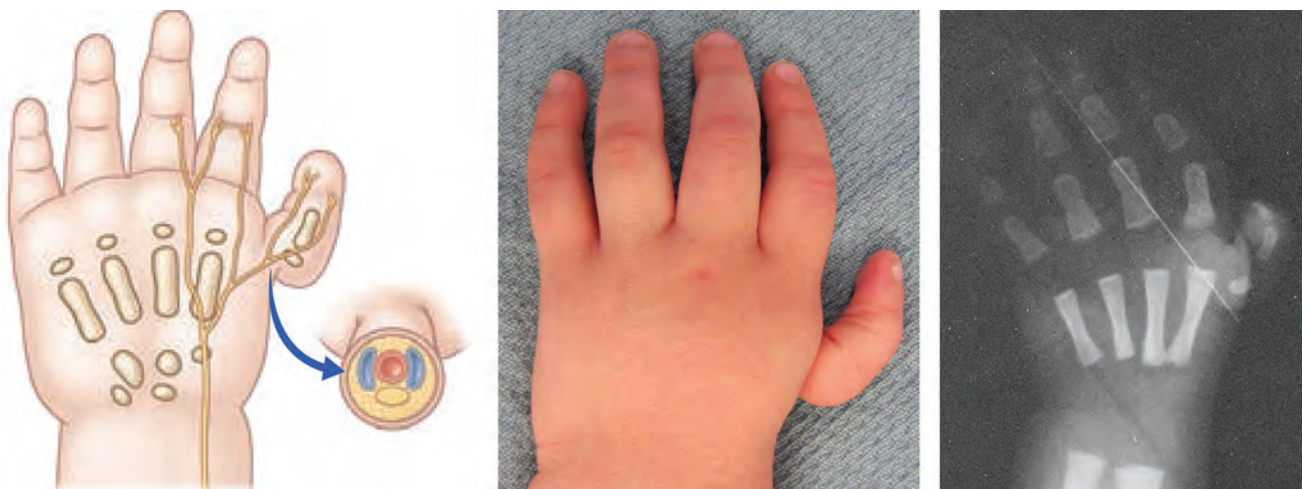


Fig. 34.5 Type IV thumb hypoplasia.

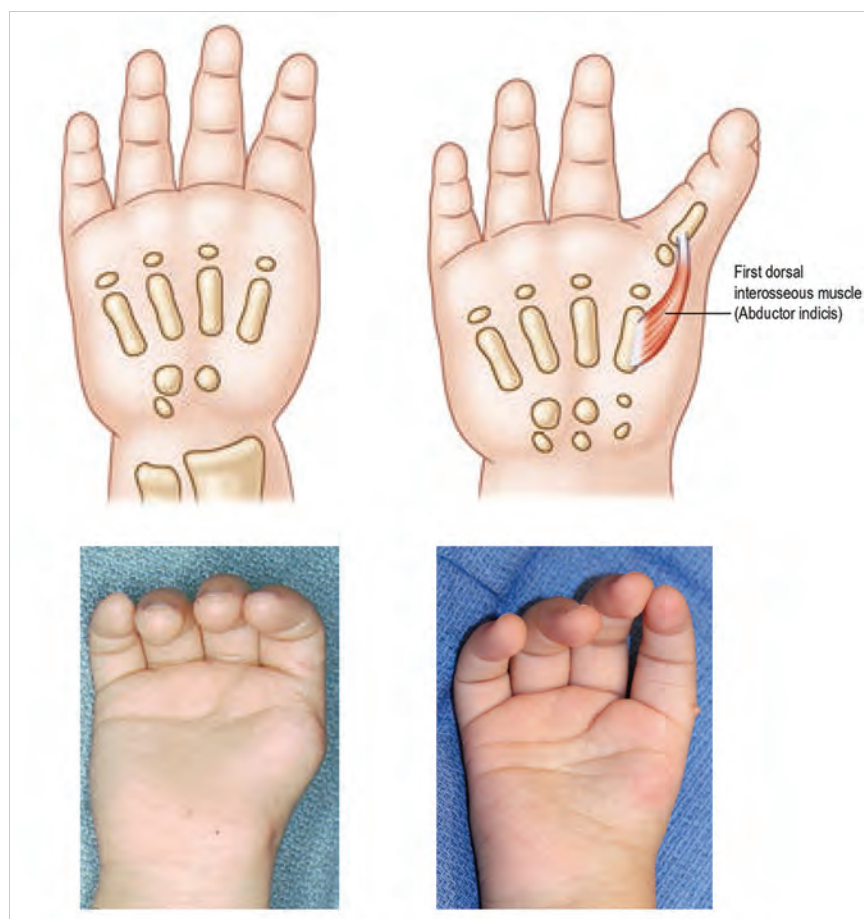


Fig. 34.6 Type V thumb hypoplasia.



Fig. 34.7 A five-fingered hand.

34.4 Operative Treatment

In the milder forms of thumb hypoplasia (types II–IIIA), surgical reconstruction addresses deficiencies in first webspace width, MP joint stability, and opposition. Treatment is tailored to the individual, depending on deficiencies in each of the aforementioned areas. In the more severe forms of hypoplasia (types IV

and V), index finger pollicization is the ideal reconstructive procedure. There is some debate in the literature about the best treatment of type IIIB thumbs. Although most authors recommend index finger pollicization, a few advocate strongly for reconstruction of the CMC joint using free vascularized metatarsophalangeal joints.

The timing for surgical intervention is debatable. For reconstruction of minor hypoplasia (types II–IIIA), most surgeons intervene around 2 years of age or older. Opponensplasty and ligament reconstruction is particularly difficult in a younger child where bone-to-bone fixation of ligament reconstruction may pose risk of injury to the growth plates. Index finger pollicization is also performed around 2 years of age when the vessels and nerves can be more easily dissected, thus making the operation safer. Although still debated, authors have shown that functional outcome after pollicization is not significantly different if the procedure is deferred until late childhood, and the outcomes of these procedures improve as children grow.

The surgical options and authors' preferred methods for reconstruction are outlined below.

34.4.1 First Webspace Deficiency

Widening of the first webspace is the most effective procedure to improve function in patients with congenital hand differences. There are many options—local transpositions flaps, local rotational flaps with or without skin grafts, regional vascular island flaps, free tissue transfer, distant pedicle flaps, and use of skin expanders. For most congenital cases, local tissue rearrangement using the four-flap Z-plasty provides the best result (► Fig. 34.8).

The contractual limb of the Z-plasty is marked first, followed by the limbs at opposing ends (transverse limbs), each at 90 degrees to the first limb. On the volar side, the transverse limb usually follows the distal palmar flexion crease. On the dorsum, the transverse limb parallels the metacarpal of the first ray. The flaps are incised and raised in the subcutaneous plane. The fascia overlying the thenar intrinsic muscles should be excised while protecting the neurovascular bundles. The flaps then “fall” into place and can be secured using 6–0 chromic sutures. Complications are rare.

For larger deficits, most surgeons prefer a dorsally based advancement flap with or without adjacent full-thickness skin grafts. Regional or free flap fasciocutaneous flaps are also quite effective.

34.4.2 Metacarpophalangeal Joint Instability

In the newborn, it is difficult to ascertain the stability of the MP joint. However, as the child gets older, it becomes obvious on

inspection. Children with weak MP joint collateral ligaments cannot grasp large objects. They will use the metacarpal head instead of the distal ray to hold objects, as everything beyond the metacarpal head is unstable. The joint should be examined in flexion and extension. Apparent laxity of the joint in extension is occasionally corrected in flexion. Often a narrow first webspace masks instability of the MP joint.

Stabilization of the MP joint can be accomplished by one of the following methods: tightening of the existing ligaments and capsule, free tendon graft reconstruction, arthrodesis or chondrodesis, or ligament reconstruction using the end of a tendon used for opponensplasty. The first method, simple imbrication and tightening, has not been effective in the long term. Arthrodesis or chondrodesis should be reserved as a last resort or in those with severe instability and poor/absent extrinsic motors. Accordingly, most surgeons choose ligament reconstruction using tendon graft.

Tendon graft can be used by itself (palmaris longus) or in the setting of simultaneous opponensplasty (flexor digitorum superficialis [FDS]). The techniques are similar. More typically, if MP joint ligament reconstruction is required, the intrinsic thenar musculature is also hypoplastic, so simultaneous opponensplasty is also chosen. The FDS tendon to the fourth digit is usually used (► Fig. 34.9). An A1 pulley incision is made in the palm and the FDS is divided as distal as possible. There can be adhesions to the fifth digit FDS or a distal bifurcation that requires a separate incision in the palm. The FDS is then retrieved through a separate incision over the distal wrist crease. It is looped around the flexor carpi ulnaris (FCU) and directed subcutaneously toward the radial aspect of the thumb MP joint. A separate slip of FCU is not used for a loop given that it causes additional adhesions. The FDS slips are separated along the fibers of the tendon until a point where it would provide maximal abduction. A transverse bone tunnel is made in the head of the metacarpal and one slip is passed through and secured with sutures—note that the growth plate of the metacarpal is proximal. A subperiosteal tunnel is made (dorsal to palmar) distal to the growth plate along the metaphysis of the proximal phalanx. The tendon is passed beneath the extensor shroud and through this subperiosteal tunnel, dorsal to palmar, and sutured to itself. Stabilization on both sides of the joint is usually needed. This technique can be modified to just use a free tendon graft if simultaneous opponensplasty is not performed.



Fig. 34.8 First webspace release with four-flap Z-plasty.



Fig. 34.9 Metacarpophalangeal joint ligament reconstruction using flexor digitorum superficialis from the ring finger. (a,b) The ligament is routed around the flexor carpi ulnaris tendon and delivered on the radial side of the metacarpophalangeal joint. (c,d) Immediate and long-term postoperative outcomes are shown.

34.5 Lack of Palmar Abduction and Opposition

Opposition transfer using one of two methods is preferred in hypoplastic thumbs: FDS tendon transfer from the fourth digit or abductor digiti minimi (ADM) transfer (with or without skin). The FDS transfer is preferred in cases where simultaneous ligament reconstruction is planned, and is usually the more commonly used method. The technique is outlined in the previous section.

In patients who need only opponensplasty (such as after index finger pollicization with poor opposition), the ADM technique is preferred. The muscle is raised through a high midlateral incision extending from the pisiform to the midaxial line of the proximal phalanx. If desired, a skin island can be taken with the muscle, thus providing the advantages of improving soft-tissue deficit and minimizing tightness of the tunnel (► Fig. 34.10). The ADM muscle is identified and its insertion is divided with a small subperiosteal extension. The sensory branch of the ulnar nerve is identified and protected. The

muscle is raised proximally to its origin at the pisiform, with care to avoid injury to its neurovascular pedicle. A radial incision over the thumb MP joint exposes the aponeurosis. The muscle is then passed through a generous subcutaneous pocket between the skin and palmar fascia and then attached to the radial aspect of the MP joint or abductor aponeurosis.

34.6 Index Finger Pollicization

Pollicization of the index finger is the ideal reconstruction for patients with types IIIB, IV, and V hypoplastic thumbs. Patients with a five-fingered hand undergo a similar procedure, a rotation–recession osteotomy, of the most radial digit.

The authors have outlined the technique for index finger pollicization in previous publications. The major goals are creation of a vascular island, rotation and recession with shortening of the metacarpal, rebalancing of the muscles and tendons, and creation of a normal-appearing first webspace. The procedure shortens the index finger and places it into a pronated, abducted position (► Fig. 34.11).

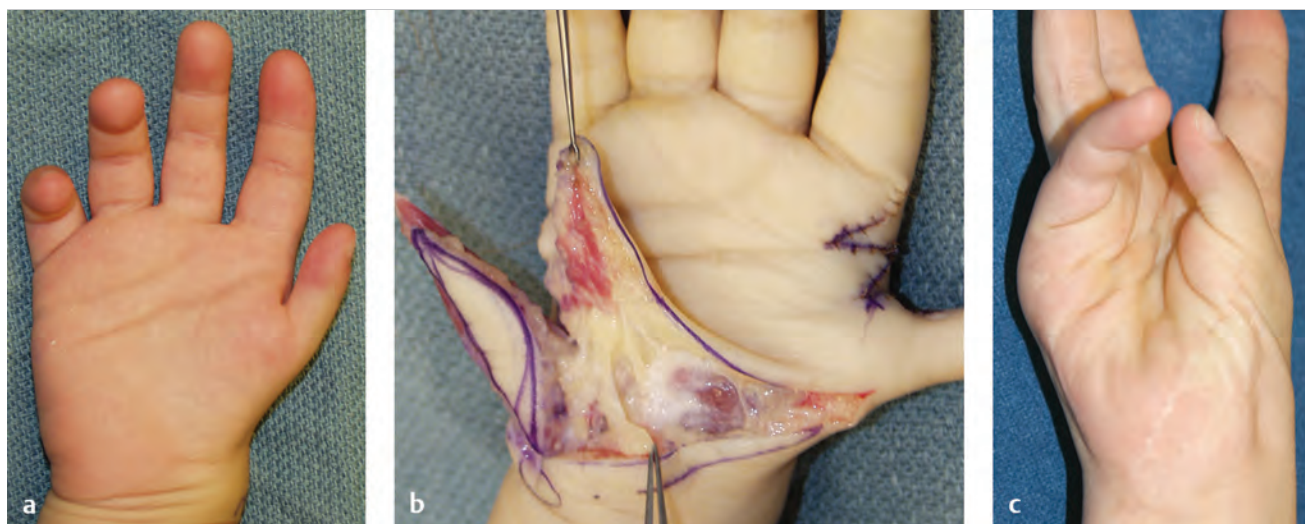


Fig. 34.10 Abductor digiti minimi myocutaneous flap transfer for palmar abduction. (a) Preoperative, (b) intraoperative, and (c) postoperative views are shown.

A racquet-shaped incision is planned around the index finger. The incision extends onto the palm where the normal thenar flexion crease would lie. An esmarch bandage exsanguinates the limb and is used as a tourniquet in the upper arm. The dorsal incision is made and two longitudinal veins can be found on either side of the extensor tendon and dissected proximally. They are located in a plane between the two layers of fat. The volar incisions are then made and the radial and ulnar neurovascular bundles are identified and dissected free. The contribution of the common digital vessel to the middle finger is ligated and divided. The intermetacarpal ligament is divided. The A1 pulley is divided. The intrinsic muscles to the index finger are mobilized off the bone with a small distal periosteal sleeve. The extra periosteum of the metacarpal is discarded to avoid ectopic bone formation. A volar oblique osteotomy is made in the metacarpal base; the retained dorsal cortex will be used to stabilize the metacarpal head, which becomes the new trapezium. The metacarpal head is then transected at the physis, and the physis is ablated. The metacarpal head is secured to the oblique-cut metacarpal base in a hyperextended position. Hyperextended placement helps to avoid future hyperextension of the joint in situ as it locks it in maximal extension in a position of function. Pins are usually not required; one or two permanent sutures will suffice. At this point, the extensors are rebalanced. The previous extensor digitorum communis is transected and sutured to the base of the proximal phalanx on its ulnar dorsal edge—this tendon will be the new abductor pollicis longus. The extensor indicis proprius tendon is shortened by the amount of metacarpal that is excised. This tendon becomes the new extensor pollicis longus. Side-to-side suturing is acceptable and works well. The intrinsic muscles are attached to bone or the extensor mechanism. Once rebalancing is complete, the neothumb should be held in neutral to slight extension, but be easily flexed with gentle pressure by the surgeon. With the new thumb held in its new position, the skin is then draped, contoured, and sutured to produce a harmonious webspace. The reconstruction is immobilized using a well-padded long arm cast for 3 to 4

weeks. Night splinting can commence for another month once the cast is removed. The flexors are not usually shortened, but in most children the flexor will shorten spontaneously over a few months.

34.7 Outcomes

Reconstructive procedures for thumb hypoplasia mimic the appearance and function of a thumb, but do not approach the range of motion, strength, and appearance of a normal thumb. Nevertheless, studies have shown that functional outcomes after reconstruction, including pollicization, are quite satisfactory. After pollicization, total active range of motion averages about 50% of normal, grip strength is 21% of normal, and use in normal activities is 84% of normal. The results are not significantly altered by age at the time of the operation, as long it is performed during childhood. Postpollicization data show that the preoperative condition of the index finger has the greatest influence on the functional outcome of the thumb. The better the status of the index finger, the better the ultimate functional outcome of the new thumb. Rigid, stiff transferred index fingers often seen in syndromic patients serve as stable posts for rudimentary grip and pinch.

34.8 Complications

Index finger pollicization is a technically demanding operation, especially when performed in small children. The structures are small, as is the margin of error. Devascularization due to injury to the arteries or, more commonly, the veins can occur. Arterial injuries should be recognized and immediately repaired. Venous problems can arise even without damage to the veins, such as by compression or kinking. If a vein is injured, it should be repaired. In the case of kinking or compression, the offending problem should be recognized and addressed. Wounds should be opened and all compression relieved.

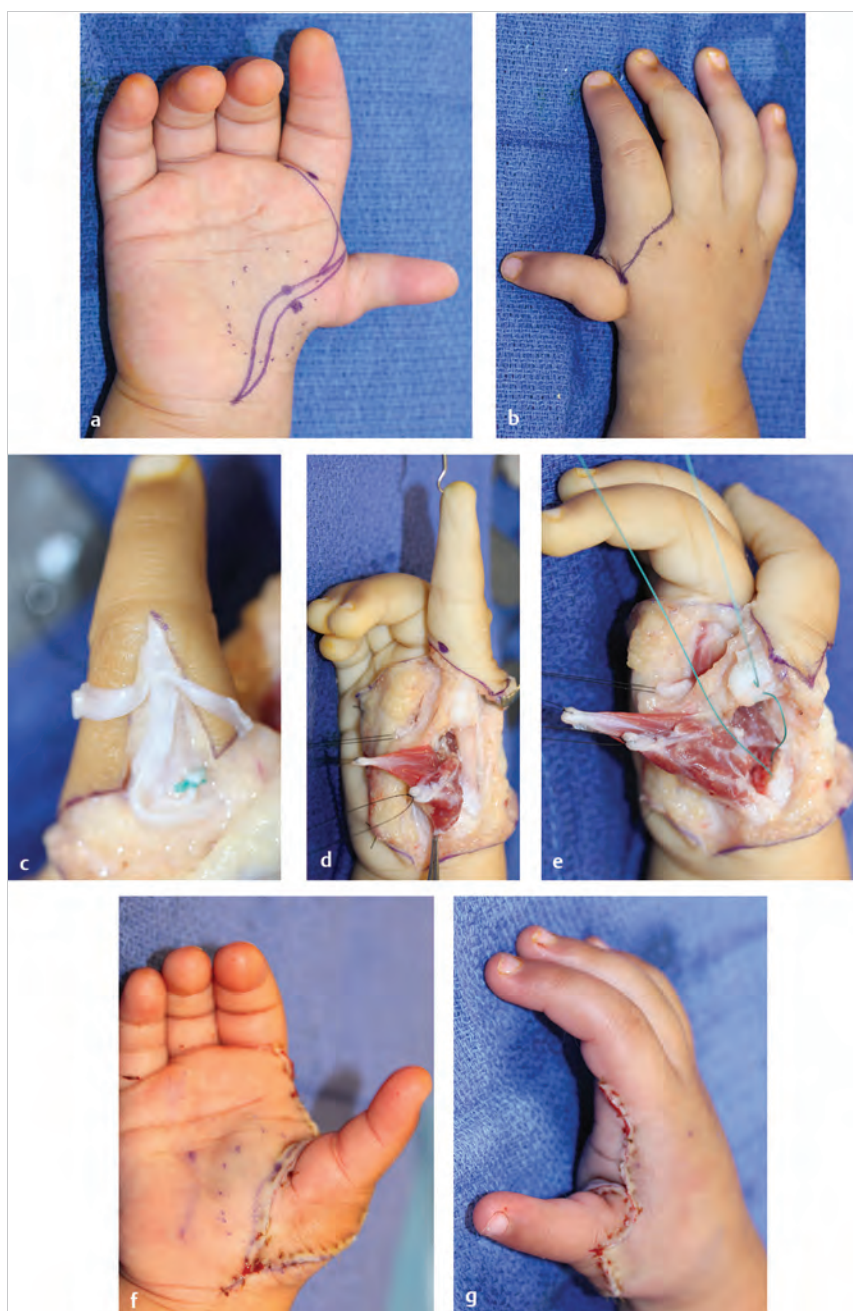


Fig. 34.11 Index finger pollicization. (a,b) show markings. (c-e) Extrinsic extensors ready to be rebalanced, intrinsic muscles and metacarpal fully exposed, and inset of the metacarpal head into the base. (f,g) Immediate postoperative outcome.

Nonunion of the metacarpal head and growth arrest of the proximal phalanx can be seen. These problems may be due to overdissection and manipulation of the blood supply. Conversely, if the growth plate of the metacarpal head is not ablated, overgrowth of the new trapezium can cause the thumb to be too long.

Skin flap necrosis can be seen with resulting contractures. These issues can be avoided with meticulous handling of the soft tissue and careful design of flaps and incisions.

By far the most common long-term problems arise from malpositioning the index finger in its new position. Very careful attention should be paid to the pronation and abduction of the new thumb to avoid the look of a short index finger in the same plane as the hand—a poor functional reconstruction.

Complications from reconstructive procedures such as first web release, collateral ligament reconstruction, and opponens-plasty are not as common. Most of these stem from poor technical execution or inadequate postoperative immobilization.

34.9 Conclusion

The thumb is the most important ray of the hand; thus, all efforts must be made to improve function in a patient with thumb hypoplasia. The Blauth classification system provides a good framework for diagnosing and treating these thumbs. Functional outcomes are satisfactory, but they provide only a fraction of the function of a normal thumb. Newer outcome

instruments are being developed that can help better quantify function after these types of reconstructive procedures.

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35 Syndactyly

Simon G. Frank and Amir H. Taghnia

Summary

Syndactyly is webbing of the digits of the hands or toes. The condition can be complete, incomplete, simple, or complex. Syndactyly usually is sporadic but may be associated with syndromes. Operative correction is performed in early childhood.

Keywords: syndactyly, pediatric, hand, feet, apert

35.1 Introduction

Syndactyly is one of the two most common congenital differences of the upper extremity, the other being polydactyly, occurring in approximately 1 in every 2,000 live births. Syndactyly is most commonly seen in isolation, though it can also be found in association with other anomalies such as Poland sequence, Apert syndrome, and the other acrocephalosyndactyly syndromes. In cases of isolated syndactyly, half of affected patients have bilateral involvement. Syndactyly is twice as common in males and there is a family history in 10 to 40% of cases, most often in an autosomal-dominant inheritance pattern with variable expression and incomplete penetrance. The feet are more frequently involved than the hands, though the majority of cases in the feet are simple, incomplete fusions of the second webspace, which often go unnoticed. In the hand, the third webspace is the most frequently affected (approximately 50%), followed by the fourth (30%), second (15%), and first (5%) webspaces. While syndactyly of the first webspace is uncommon, it tends to be associated with syndromes and part of a highly complex syndactyly of the hand.

Syndactyly is typically classified as either *complete*, in which the webbing extends to the tip of the digit and involves the nail complex, or *incomplete*, in which the webspace begins distal to the level of the normal commissure but does not extend to the tip (► Fig. 35.1). Distinction is next made on the basis of the tissues forming the interdigital connection; a *simple* syndactyly involves only skin and abnormal fibrous tissues, whereas a *complex* syndactyly includes abnormal osseous or cartilaginous connections. Finally, a syndactyly is termed *complicated* when the connections are more than simple side-to-side fusions. The

Apert hand, central polysyndactyly, and typical cleft hand are examples featuring complicated syndactyly.

The etiology of syndactyly remains unclear. Simple syndactyly can be explained by the failure of programmed cell death within the webspace, which forms the basis of the traditional classification of syndactyly as a failure of differentiation under the original Swanson/IFSSH (International Federation of Societies for Surgery of the Hand) system. However, more complex forms of syndactyly lack a well-accepted cause and there is no current consensus on their appropriate classification. Recently, the IFSSH has proposed adopting the OMT (Oberg–Manske–Tonkin) classification, which categorizes syndactyly in the “abnormal axis formation/differentiation–hand plate” subcategory within the larger category of malformations.

35.2 Diagnosis

The diagnosis of syndactyly is made by physical examination. The involved digits, the extent of the syndactyly, and the involvement of the nail bed should be noted. The distinction between a simple and complex syndactyly can often be made based on the relative motion of the adjacent digits. Examination should also be performed of the remainder of the upper extremity, the contralateral upper extremity, and the feet. Radiographs of the affected hand should be obtained in most cases, but are generally unnecessary in the case of simple syndactyly. Imaging by ultrasound, magnetic resonance imaging (MRI), or angiography may be helpful in the most complex cases to determine soft tissue and vascular anatomy, or to describe the bony configuration when bone overlap precludes accurate determination by plain radiographs. The authors do not perform routine angiography, MRI, or ultrasound, however. Further workup or referral to other specialists may be indicated in syndromic cases of syndactyly, though these patients are often already involved in a multidisciplinary care team.

35.2.1 Nonoperative Management

There is a limited role for conservative management in syndactyly. Surgical intervention may not be indicated for minor forms

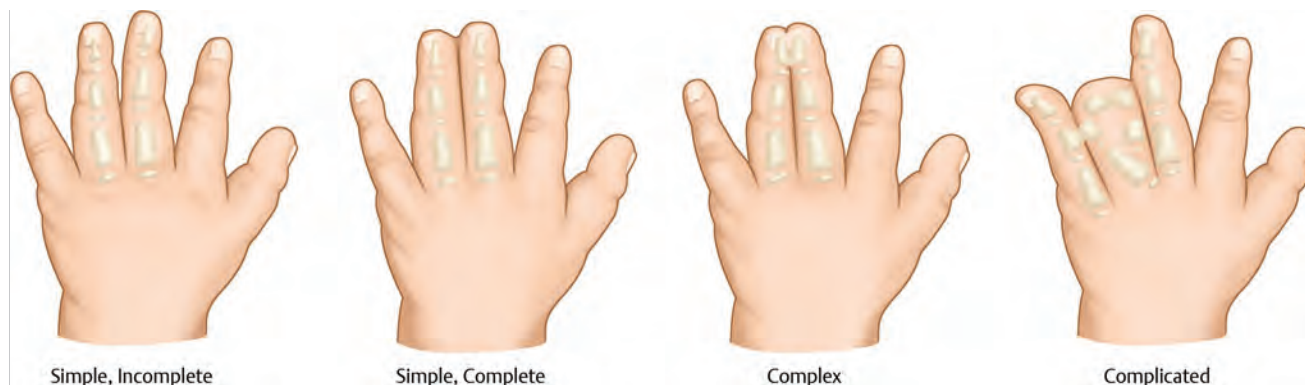


Fig. 35.1 Classification of syndactyly.

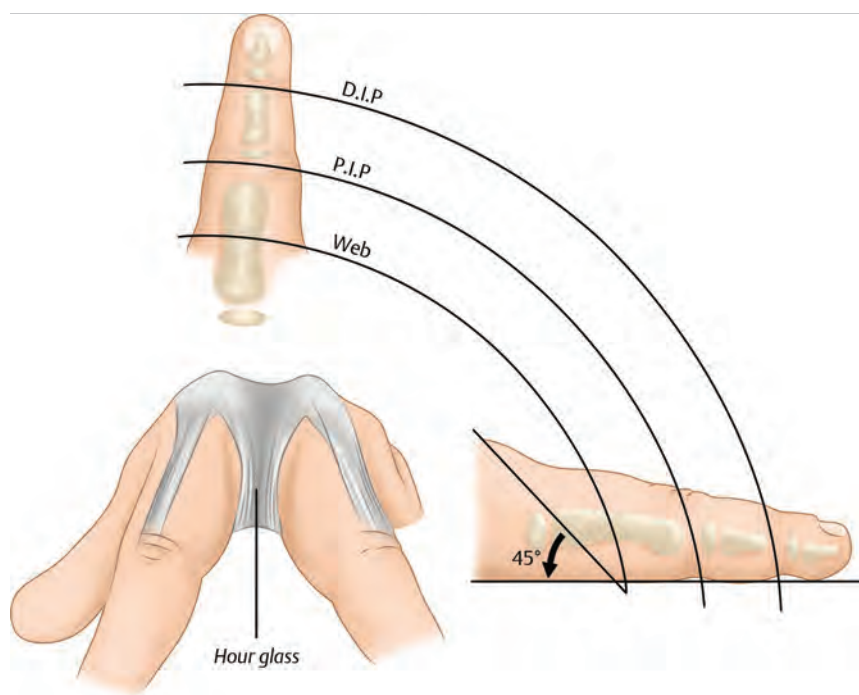


Fig. 35.2 Illustration of the 45-degree inclination and hourglass configuration of the normal web.

of syndactyly in which there is no functional impairment, or it may be at the discretion of the parents. Particularly in the feet, incomplete syndactyly can be relatively inconspicuous, and correction is therefore often unnecessary. Medical tattooing has been proposed to simulate a webspace in the toes, and has even been embraced by some patients in a creative manner.

35.2.2 Operative Treatment

Anatomy

Prior to any surgical correction of syndactyly, it is necessary to understand the normal anatomy of the webspace (► Fig. 35.2). The normal interdigital webspace has an hourglass shape, and is inclined 45 degrees from dorsal to palmar. The second and fourth webspaces are wider than the third webspace, consistent with the greater abduction possible with the index and small fingers. The normal position of the base of the commissure is the midportion of the proximal phalanx (when the fingers are maximally splayed apart), which is approximately halfway between the distal palmar crease and the proximal interphalangeal joint crease. These landmarks place the second and third webspaces at the same level, with the fourth lying slightly proximal. The first webspace “has the configuration of a diamond-shaped tetrahedron,” with triangles of glabrous skin volarly and nonglabrous skin dorsally. The edge of the web extends as a smooth arc from the level of the index metacarpophalangeal joint to a point just distal to the thumb metacarpophalangeal joint.

The abnormalities present in syndactyly can be highly variable. In all cases, there is a deficiency of skin, which can be demonstrated to parents by comparing the measurement of the circumference of two digits held together versus the sum of the circumferences of the individual digits. The deficiency is particularly pronounced in the area of the normal commissure,

explaining the need for skin grafts in this area with most releases. The fused digits are connected in the midaxial line along the length of fusion by an abnormal fascia. The digital nerves and arteries are often abnormal with regard to their branching patterns, most frequently bifurcating more distally than normal, especially in more severely affected cases. This distal bifurcation may limit the extent of separation of the digits or may (rarely) necessitate ligation of one of the digital arteries and intraneural separation of the digital nerves. In complex syndactylies, the skeletal union is usually at the level of the distal phalanx, but can be along the entire digit in some cases. Finally, in complicated syndactylies, the anatomy can be highly abnormal, including duplications within the web (polysyndactyly), transversely oriented skeletal elements, joint abnormalities, and abnormalities of the phalanges, to the point where recreation of a normal, useful digit is impossible.

Indications

As already noted, surgical treatment is the mainstay for syndactyly. Exceptions include the aforementioned minor cases of syndactyly with no functional impairment, particularly in the feet, as well as highly complex syndactylies in which surgery may in fact worsen function. Severe medical comorbidities can also present a contraindication to surgery.

Timing of Surgery

Historically, release of syndactyly has been performed anytime from the neonatal period to well into childhood. Children establish patterns of prehensile function by 24 months of age, which many feel serves as an upper limit for completing releases. The timing of surgery is often influenced by how much surgery is needed—since most surgeons advocate against releasing neighboring webspaces simultaneously, patients with multiple

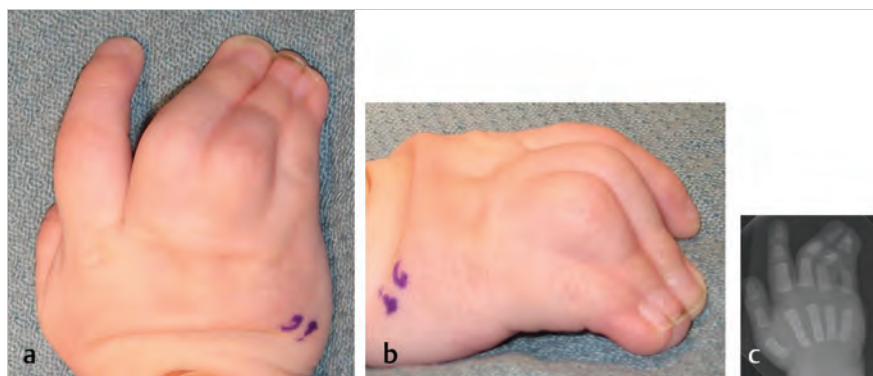


Fig. 35.3 Demonstration of angular deformity due to complete complex syndactyly involving the fourth webspace. (a) Dorsal view. (b) Oblique view. (c) Radiographic view.



Fig. 35.4 Four-flap Z-plasty for deepening of the first webspace. (a) Preoperative markings. (b) Intraoperative view after elevation of flaps. (c) Final view showing inset of flaps.

syndactylies require multiple operations to complete the entire release. Neonatal surgery has been advocated due to the abundance of mobile skin and thus the ability to avoid skin grafts; however, this is not routinely performed by most surgeons. There remains no consensus for ideal timing of surgery, though most surgeons favor performing releases between 9 months and 2 years of age. In most cases, there is no harm in waiting until the child is older, though parents are often eager to have releases performed sooner. Certain cases, however, warrant earlier intervention, specifically release of the first and fourth webspaces, as well as complex cases in which multiple surgeries are planned. In the case of complete syndactylies of the first or fourth webspace (especially in cases where distal bony coalition is present), surgery is best performed early to avoid progressive skeletal deformity and flexion contractures, and to allow for the development of pinch in the case of the first webspace (► Fig. 35.3). In complex cases, the first of multiple surgeries may need to commence earlier than normal to allow for completion of all releases within an appropriate time frame, as well as to allow for possible intervening procedures by other specialists in the case of children with associated anomalies.

Surgical Principles

A variety of techniques have been described in the history of syndactyly release. Successful techniques will generally adhere to a number of principles, though there are always exceptions. Surgery should only be performed on one side of the digit at a time to avoid vascular compromise to the digit or the skin flaps. The commissure should be resurfaced with some type of skin

flap. Zigzag incisions should be used volarly to avoid flexion contractures. When needed, skin grafts should be full thickness. Finally, care must be taken with postoperative dressings and immobilization.

Release of Incomplete Syndactyly

In situations in which only mild deepening of the webspace is required, a variety of local flaps have been described. The majority of proposed repairs involve some manner of double-opposing Z-plasties, as in the “jumping-man” flap. As the syndactyly begins to approach the proximal interphalangeal joint, one should consider a formal syndactyly release.

Release of First Webspace

As previously mentioned, syndactyly of the first webspace is more common in syndromic cases, such as Apert syndrome. Reconstruction of this unique webspace can also be considered in the context of hypoplastic thumb as well as trauma, and many of the same techniques can be applied to syndactyly.

For minor deficiency of the first webspace, a variety of Z-plasties have been described. A standard single Z-plasty with large flaps may be sufficient, but the four-flap Z-plasty is ideally suited to this situation, and provides good length and contour of the webspace (► Fig. 35.4).

With more significant deficiency, tissue must be introduced into the webspace to provide adequate abduction and grasp. Many local flaps have been described, both with and without the use of skin grafts, including transposition flaps from the

dorsum of the thumb or index finger, rotation or transposition flaps from the dorsum of the hand, and V-Y advancement flaps. An advancement flap of the dorsal skin into the defect is an effective technique. The necessary skin grafts should be placed along the adjacent borders of the thumb or index finger rather than across the webspace. This technique takes advantage of the mobile dorsal skin and allows repeated advancement of the tissue if required.

Regional flaps, such as the reverse radial forearm flap or reverse posterior interosseous artery flap, are sometimes needed in severe cases such as the typical cleft hand, certain symbrachydactylies, and the mitten hand. Though these regional options provide ample skin, they can be bulky in a young child due to the thickness of the adipose layer, and they also leave the child with a potentially unsightly donor site on the forearm. Free flaps may also be used, and have been described from the lateral arm, groin, and contralateral forearm.

In addition to adequate soft-tissue coverage, release must also be performed of fibrous bands between the thenar muscles, and possibly of the carpometacarpal joint. The vascular anatomy is frequently abnormal; therefore, care should be taken during the dissection, and division of the branch of the common digital artery leading to the index finger must occasionally be performed provided the index finger has adequate circulation from the ulnar side. Finally, it is occasionally preferable to perform a ray resection of the index finger as long as three functional fingers remain.

Release of Complete Simple Syndactyly

A multitude of techniques have been described for the release of the complete simple syndactyly. Conceptually, the operation can be thought of as three components: the flap used to line the commissure, the digital flaps and skin grafts used to line the fingers, and the reconstruction of the nail folds. Nail fold reconstruction will be discussed in the following section.

The majority of current techniques use a dorsally based rectangular (or hourglass-shaped) flap to line the commissure, though a combination of dorsal and volar triangular flaps can also be used. Rectangular flaps theoretically best approximate

the configuration of the normal commissure, while interdigitating triangular flaps allow greater flexibility in their inset, which may be of value in more complex cases where one may not be able to achieve the full depth of the web due to anatomic abnormalities. Lateral tabs can also be added to the rectangular flaps, which when rotated down into the commissure can help line the lateral aspect of the digits, reducing or eliminating the need for skin grafts at the expense of a scar on the dorsum of the hand and the potential for tighter closure.

Flaps to line the digits are typically made using zigzag incisions dorsally and volarly, though some incorporate a volar rectangular flap proximally to reline the lateral aspect of the digit adjacent to the commissure, while still other techniques make use of straight-line incisions dorsally. With standard zigzag incision, techniques vary in the acuity of the angles for the flaps: more acute angles will ensure there are no straight-line scars along the digit that could lead to contracture, while more oblique angles will provide better vascularity to the tips of the flaps.

Finally, when skin grafts are required, full-thickness skin is preferable for its decreased contraction and improved appearance. A number of potential sources for skin have been described. The hypothenar eminence provides skin from the same operative field, though it can be thick and nonpliable. Skin can be obtained from various sites on the arm including the volar wrist crease, the antecubital fossa, and the medial upper arm, though all these leave potentially visible scars. The foreskin can be used at the time of circumcision, and though this skin is otherwise discarded, using it as a graft can result in undesirable pigmentation. Finally, the inguinal crease or lower abdomen can provide large amounts of skin and leave an inconspicuous scar, for which reasons many authors consider it an ideal donor site provided the future hair-bearing skin is avoided. Though there is an appeal to avoiding skin grafting, the use of a skin graft on the lateral aspect of the digit in fact offers few downsides and should be used without hesitation if there is concern regarding an overly tight closure.

The authors' preferred technique, consisting of a dorsal hourglass-shaped flap and dorsal and volar zigzag incisions, will be described (► Fig. 35.5). Markings are made of the dorsal skin dimples, denoting the metacarpophalangeal joints. These points

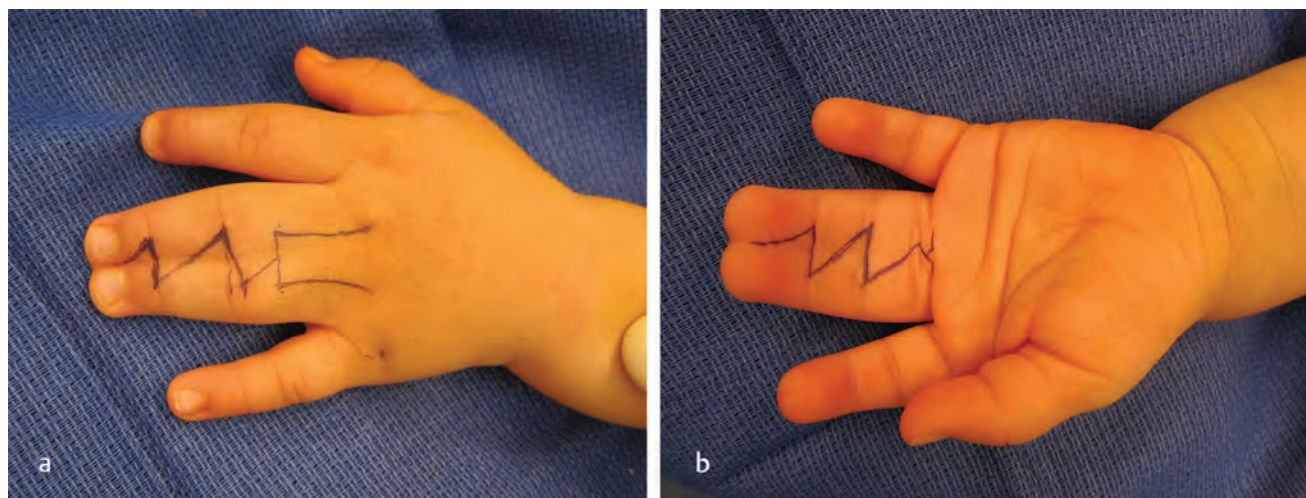


Fig. 35.5 Markings for complete simple syndactyly release. (a) Dorsal view. (b) Volar view.

form the base of the hourglass-shaped flap. The distal extent of the flap is marked a few millimeters distal to the planned level of the commissure to allow for loss in length of the flap as it is rotated volarly. Laterally, the flap extends to the midline of the digits. The level of the commissure can be determined by a number of methods. Comparison is made to the adjacent webspaces, recalling that the fourth webspace lies slightly proximal. The contralateral hand can be examined and measurements taken. Finally, the commissure should lie at the midportion of the proximal phalanx when the fingers are maximally splayed apart. This point is approximately halfway between the distal palmar crease and the proximal interphalangeal joint crease.

Zigzag incisions are then marked on the dorsum of the digits, with lines connecting the adjacent proximal interphalangeal joints and similarly the adjacent distal interphalangeal joints. Markings are then completed to form the triangular flaps, taking into consideration which orientation of markings will yield an appropriately angled set of flaps. The markings extend laterally to the midpoint of each digit.

Attention is then turned volarly. A transverse line is marked volarly approximately a millimeter proximal to the level of the planned commissure to allow for creep, extending laterally to the midline of each digit. A small triangular flap is added at the midpoint of this line, which will serve to break up the linear scar along the base of the commissure. This triangular flap will be inset into a small longitudinal incision made into the rectangular flap at the time of closure. A set of zigzag markings that matches the dorsal markings is then made. Volar and dorsal markings should be confirmed to ensure the flaps will inset appropriately once raised.

After exsanguination and inflation of the tourniquet, the rectangular flap is incised and raised in the subcutaneous plane. The web is released, taking care to avoid damaging the mirroring flaps during incision, particularly distally where the webbing may be thin with minimal intervening soft tissue. Flaps are raised to the midline. The digits are held in abduction, and the neurovascular bundles are identified as they approach the commissure. The interdigital fascia is incised to allow release of the digits, and preferably excised, particularly if thick and fibrous.

Dissection continues as far proximally as required or as dictated by the branching of the neurovascular structures. Intra-neural dissection and ligation of an arterial branch may be required, as long as each digit retains a digital artery on at least one side. If there is any doubt about the vascular anatomy, adequate circulation should be confirmed by applying a microvascular clamp to the branch to be divided, deflating the tourniquet and verifying that all digits remain well perfused. Conservative defatting of the digit may be performed if necessary for closure, taking care to protect the neurovascular bundles.

If present, synostoses may be divided with a knife or small osteotome in younger children, while an oscillating saw may be required for older children. Duplicated skeletal components may be excised and collateral ligaments re-inserted. Where necessary, corrective osteotomies are generally deferred to a second stage when the child is older.

Flaps are inset, taking care to avoid excess tension. A small releasing incision is made at the midpoint of the dorsal rectangular flap to accept the volar triangular flap. Absorbable sutures such as 5-0 or 6-0 chromic gut are generally preferable in the younger patient. After inset is complete, the tourniquet is deflated and hemostasis obtained with bipolar cautery. Templates are made of open areas using the foil from a suture package, and full-thickness grafts are harvested, defatted, and inset (► Fig. 35.6). Block of the digits may be performed at this point using 0.25% plain bupivacaine.

Application of an appropriate dressing is critical. An antibiotic-impregnated nonadherent gauze is applied to the incisions and grafts, followed by lightly moistened gauze and then an adequate amount of moistened cotton to stent the webspace. A bulky dry gauze dressing is then secured using Kling wrap. The extremity is immobilized in a well-padded long-arm cast with the elbow in flexion. The cast should be left on for 3 weeks.

Once the cast is removed, the parents are encouraged to wash the extremity in the bathtub and rub the incision areas with a moist washcloth. A hand therapist will make a splint with a silicone webspace conformer to stent the webspace open and put pressure on the incisions. This splint/conformer should be worn at night for 1 month after the cast is removed.

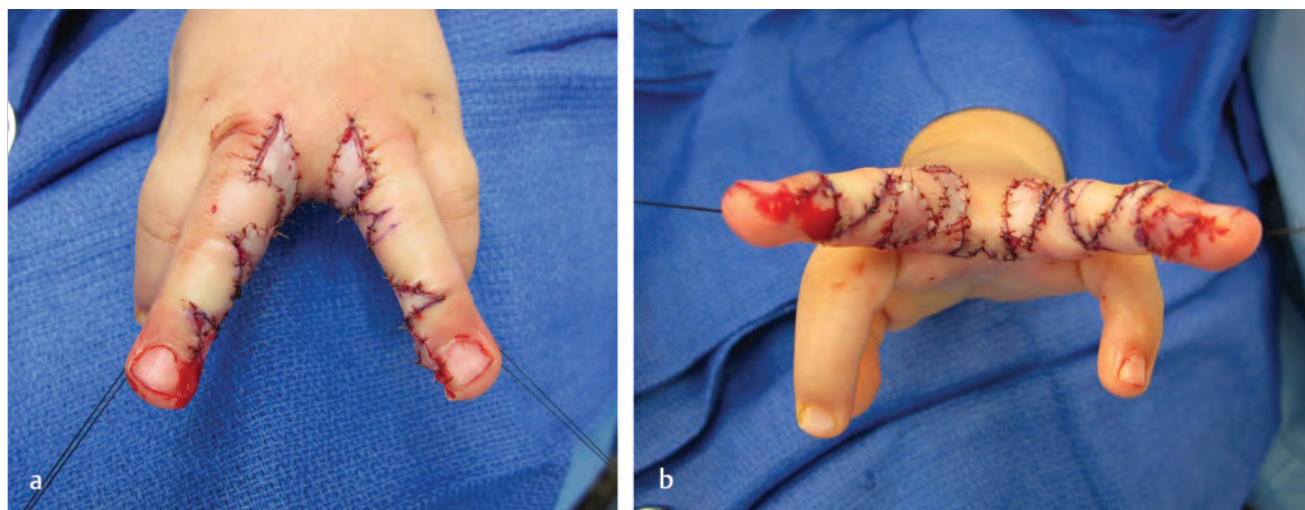


Fig. 35.6 Complete simple syndactyly release following inset of flaps and skin grafts. (a) Dorsal view. (b) View of commissure demonstrating inset of small volar triangular flap into releasing incision on distal aspect of hourglass-shaped flap.

Reconstruction of Nail Folds

Successful reconstruction of the nail fold remains a challenge, for which a variety of techniques have been described. Prior to reconstruction of the folds, it is important to trim the nail matrices and underlying bone down to an appropriate width. Following this step, for minor deficiencies the lateral skin can be undermined and simply advanced. Alternatively, release of the fingertip can be performed asymmetrically, covering one side with local skin and the other with a full-thickness skin graft. Composite grafts of skin and pulp from the toe may also be used. Finally, the nail fold flaps of Buck–Gramcko are effective and are preferred by many surgeons. Ultimately many of the described techniques result in ischemic flaps of questionable utility, and if used, flaps should therefore be fairly wide.

Apert Syndrome

The hand deformity seen in Apert syndrome consists of a complex syndactyly of the index, long and ring fingers joined by a simple syndactyly to the small finger. The thumb is short, is variably joined to the central mass, and may have a radial clinodactyly. There is frequently a synostosis of the base of the ring and small finger metacarpals. There are also typically deformities of the shoulder and elbow, though these are rarely treated

surgically. Upton has classified the Apert hand into three categories (► Table 35.1). The unique abnormalities seen in the syndrome dictate certain modifications to standard syndactyly releases. The symphalangism and resultant lack of interphalangeal joint motion in the central three digits allows for straight-line releases along the digits without concern for flexion contracture (► Fig. 35.7). Because of the potential for neurovascular abnormalities within the webspaces, interdigitating triangular flaps are generally used for commissure reconstruction to allow for greater flexibility in setting the depth of the web. When covering the lateral aspect of the digits, many surgeons prefer to leave distal areas open and allow them to epithelialize, while alternatively full-thickness grafts may be applied directly over exposed bone with generally satisfactory results. Regardless of preference in this regard, requirements for skin graft are generally large, making the lower abdominal donor site ideal, particularly when performing bilateral releases as is often the case in Apert syndrome. Finally, the complexity of the Apert hand necessitates a staged approach, and the surgeon should not hesitate to offer revisions as the child grows.

Many treatment algorithms have been proposed, which vary primarily in how the digital separations are staged, as well as timing of the operations. Irrespective of which algorithm is followed, the timing of surgeries will need to be coordinated with the craniofacial team.

Table 35.1 Classification of the Apert hand

	Type I (“obstetrician’s hand” or “spade hand”)	Type II (“cup hand” or “mitten hand”)	Type III (“rosebud hand” or “hoof hand”)
Thumb	Free; radial clinodactyly	Joined by simple syndactyly; radial clinodactyly	Joined by tight complex syndactyly; radial clinodactyly may be absent
Central mass	Flat plane; complex syndactyly	Cupped transverse arch with splaying of digits at metacarpophalangeal level; complex syndactyly	Tightly cupped transverse arch with incorporation of thumb; complex syndactyly
Fifth digit	Joined by simple syndactyly, typically incomplete	Joined by simple syndactyly, typically complete	Joined by complex syndactyly



Fig. 35.7 Markings for release of syndactyly in an Apert hand. Note use of straight-line incisions and triangular flaps. (a) Volar view. (b) Dorsal view.

Release of the thumb webspace is critical and should be done early. This can be accomplished with the standard techniques already described, bearing in mind that re-advancement will likely be required at a future stage. In severe type III hands, there may be limited tissue available to advance into the webspace, in which case free tissue transfer can be considered. Alternatively, skin grafts can initially be used on the sides of the web followed by successive re-advancement of the dorsal flap. Finally, the type III Apert hand represents one of the situations in which sacrifice of the index ray may be preferable in order to reconstruct an adequate web. Whichever technique is used, the eventual lengthening of the thumb phalanx at the time of clinodactyly correction will improve the webspace.

At the time of thumb webspace release in type III hands, the tightly bunched digital mass can be converted to the flat configuration of a type I hand by performing distal osteotomies of the digital synostoses through a hyponychial incision. The digits are then held in a more collinear arrangement with a single transverse K-wire. This procedure makes subsequent releases much more straightforward, and also improves the maceration of the nail beds. In type I or II hands where this procedure is not required, the fourth webspace syndactyly is generally released at this time to preserve mobility and prevent further deformity to the fifth digit.

Staged release of the remaining digital syndactylies is then performed, with the goal of completing the releases well before school age. As with the first stage procedures, these surgeries should be performed bilaterally. A two-team approach can be helpful to minimize anesthetic time.

Despite best efforts in correcting these hands, they will never look nor function normally. Additionally, they will inevitably require revisions, with rates approaching 100%. Web creep can be particularly pronounced in the second web due to the additional epiphysis at the metacarpal base. Some surgeons therefore prefer a dorsal rectangular flap in this webspace given it can be re-advanced as needed. Other potential revisions include repeat osteotomy of the synostosis between the fourth and fifth metacarpals, as it often reforms. Various techniques have been proposed to limit this recurrence including the use of interposition fascia, fat, or cadaveric dermis grafts.

Poland Sequence

In addition to the chest wall deformities discussed elsewhere in this book, the hand in Poland sequence can be hypoplastic with variable symbrachydactyly. The central digits are more frequently affected. Treatment is customized to the deformity and should follow standard principles for syndactyly release, though many surgeons prefer to exaggerate the depth of the webspace in patients with short digits, possibly including a release of the intermetacarpal ligament. Hand surgeries should be coordinated with the rest of the patient's treatment plan.

35.3 Complication

Early complications from syndactyly release include infection, graft loss, and flap necrosis, all of which can generally be managed without further surgery. Many of these complications can be avoided with adequate postoperative immobilization, which is particularly crucial to avoiding graft loss. Large areas of graft

loss necessitating regrafting are rare, though regrafting should be considered especially on the volar surface or close to the depth of the web. Rarely, adhesion of the fingers can occur, requiring a repeat release.

Late complications include web creep, scarring, deviation of the digits, joint instability, and complications related to skin grafts such as pigmentation or hair growth. Re-deepening of webspaces, particularly in the case of the first webspace, may be required. This complication may be partially avoided by thorough release of the fascial bands within the first webspace at the time of initial surgery. Appropriate flap selection in cases where recurrence is expected, as in Apert syndrome, allows for re-advancement of the flaps. In all cases, formation of scar contracture or hypertrophic scars may necessitate excision and closure by skin grafting and re-advancement of flaps where possible. Rotational and angular deformities are more common after complex syndactylies, and may require osteotomies.

35.4 Conclusion

The surgeon should master a few effective techniques for treating syndactyly and use them consistently. Once mastered, those techniques can then be modified based on experience. Regardless of which techniques are used, immobilization is critical to avoiding complications. Following surgery, patients should be followed closely as they grow so that any problems with web creep can be addressed early. Additionally, cases should be carefully documented with photographs. The combination of thorough documentation and ongoing observation of the patient allows for the "if/then" feedback necessary for the surgeon to learn and improve.

35.5 Key Points

- Border digits should be released early to avoid progressive deformity and ensure optimal hand function.
- During separation of syndactyly, care must be taken to avoid damage to the neurovascular structures, which become increasingly anomalous as the complexity of the syndactyly increases.
- Adequate postoperative immobilization is critical to obtaining satisfactory outcomes.
- Management of syndactyly in Apert syndrome requires a specialized approach, with differences in timing, goals, and surgical technique.

Suggested Readings

- Fearon JA. Treatment of the hands and feet in Apert syndrome: an evolution in management. *Plast Reconstr Surg*. 2003; 112(1):1–12, discussion 13–19
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36 Polydactyly

Paige M. Fox, Heather L. Baltzer, and Steven L. Moran

Summary

Radial, central, and ulnar polydactyly represents the spectrum of digital duplications seen in pediatric patients. From soft-tissue-only ulnar polydactyly to complex central polydactyly, the diagnosis, management, outcomes, and complications of each duplication type are discussed. Classification systems along with their inherent strengths and weaknesses are included. Multiple clinical and radiographic examples are provided. After reading this chapter, the clinician will feel more comfortable recognizing, diagnosing, and treating the wide spectrum of digital duplications.

Keywords: radial polydactyly, ulnar polydactyly, preaxial polydactyly, postaxial polydactyly, duplication, triphalangeal

36.1 Introduction

Polydactyly, classified as a duplication of parts, is the most common congenital hand anomaly. Polydactyly involving the thumb is referred to as radial or *preaxial polydactyly* and duplication that involves the small finger is referred to as ulnar or *postaxial polydactyly*. Ulnar polydactyly is more common in African races, while radial duplication is more common in the Caucasian and Asian races. *Central polydactyly*, duplication of the nonborder digits, is less common than preaxial or postaxial polydactyly. Central duplication is often associated with syndactyly and cleft hand. Its treatment is, therefore, more complex, requiring treatment of the polydactyly, the syndactyly, the central cleft, and any accompanying webspace tightness.

36.1.1 Radial Polydactyly

Radial polydactyly has an incidence of 1 in every 3,000 births. Variable degrees of duplication exist and range from widening of the thumb tip to complete duplication. The most common classification of thumb duplication has been described by Wassel, with type IV representing the most frequent pattern. Types I, III, and IV refer to bifid phalanges, while types II, IV, and VI refer to complete phalangeal duplication. Wassel type VII describes a thumb with an extra, or third, phalanx, also known as a triphalangeal thumb (► Fig. 36.1). Radial polydactyly with triphalangism is associated with an autosomal-dominant inheritance pattern and genetic mutations within the hedgehog pathway. The remaining types of preaxial polydactyly are thought to be the result of spontaneous mutations. Radial polydactyly often occurs in isolation but may be associated with Fanconi anemia or Holt–Oram syndrome.

36.1.2 Central Polydactyly

Central polydactyly (duplication of the index, middle, or ring fingers) is often seen in combination with other anomalies, most commonly syndactyly, known as synpolydactyly. This is felt to be a heritable condition with a broad spectrum of

phenotypic variability. Stelling and Turek have classified central polydactyly into three main types: (1) extra soft-tissue mass only, (2) duplication of a digit or partial digit that articulates with an enlarged or bifid phalanx or metacarpal, and (3) duplication of a digit including the metacarpal and all soft tissues involved (► Fig. 36.2). In 2015, Wall et al reviewed 56 hands in 40 patients to establish a radiographic classification for synpolydactyly. The system is based on level of duplication with type I representing metacarpal duplication and type III representing middle or distal phalanx duplication (► Fig. 36.3).

36.1.3 Ulnar Polydactyly

The original ulnar polydactyly classification by Stelling and Turek was a three-part classification system similar to that described earlier for central polydactyly. More commonly, ulnar polydactyly is classified into types A and B as described by Temtamy and McKusick. Type A represents a well-formed extra digit that articulates with the small finger metacarpal or a duplicate small finger metacarpal (► Fig. 36.4). Type B ulnar polydactyly describes a rudimentary digit with only soft-tissue attachments to the small finger (► Fig. 36.5). Type B can present as a spectrum ranging from a small nubbin on the ulnar border of the digit to a relatively well-formed digit with only soft-tissue attachment and no articulation. The presence of a nubbin is thought to represent a remnant or neuroma of a pedunculated supernumerary digit that autoamputated in utero. Pritsch et al have proposed a subclassification of type A ulnar polydactyly: type 1, fully developed sixth metacarpal; type 2, intercalated supernumerary digit between ring and small finger with rudimentary metacarpal; type 3, supernumerary digit ulnar to small finger with rudimentary metacarpal; type 4, digit articulates with fifth metacarpophalangeal joint; and type 5, supernumerary digit arises from a bifid proximal phalanx. Patients with type A ulnar polydactyly present with bilateral hand involvement quite commonly (69%) and also frequently have foot involvement. The majority of type B ulnar polydactyly is bilateral.

Type B postaxial polydactyly is uncommon in Caucasian infants, but relatively common in African American infants, with respective prevalence of approximately 1 in 1,300 and 1 in 150 live births. Type A is relatively rare and has been reported as equal between African American and Caucasian patients, but a recent review demonstrated increased relative frequency among Caucasians and Hispanic patients compared with African American patients. Isolated, nonsyndromic ulnar polydactyly, inheritance is more commonly an autosomal-dominant pattern, but recessive patterns of inheritance have also been described. Associated congenital anomalies and syndromes can also be present with ulnar polydactyly including Ellis–van Creveld, Smith–Lemhi–Optic, McKusick–Kaufmann syndromes, short-rib polydactyly syndrome I, orofaciocigital syndrome III, Bardet–Biedl syndrome, Meckel–Gruber syndrome, Greig cephalopolysyndactyly, and Pallister–Hall syndrome.

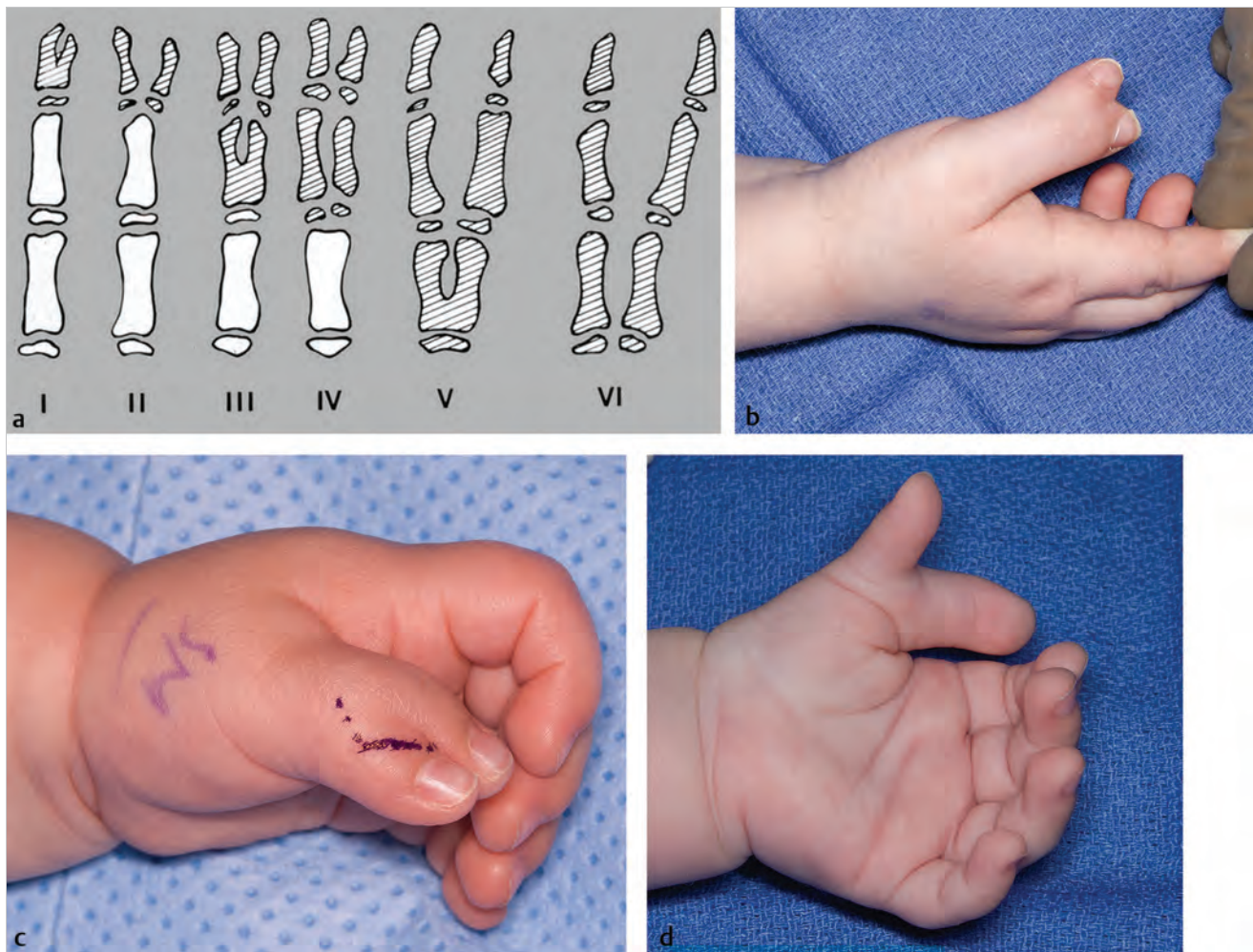


Fig. 36.1 (a) Diagram of the Wassel classification scheme for thumb polydactyly. (b) Clinical example of a Wassel type II thumb polydactyly. (c) Clinical example of type III thumb polydactyly. (d) Clinical example of type IV thumb polydactyly.

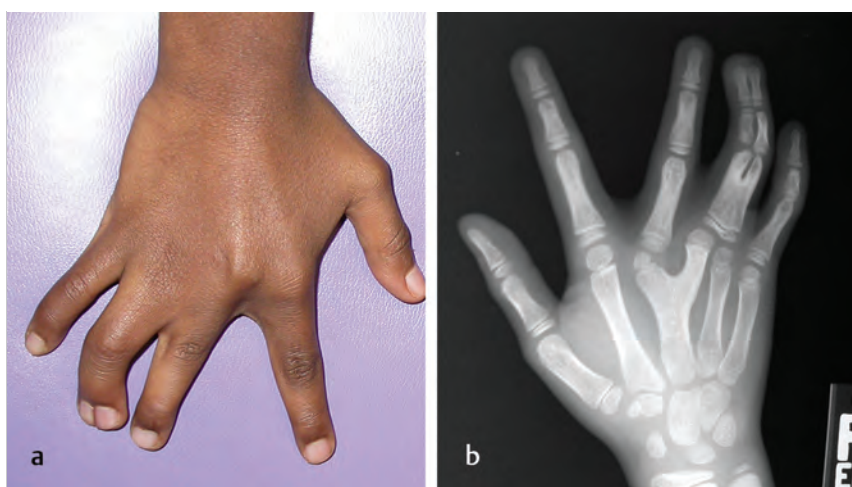


Fig. 36.2 (a) Clinical example of central polydactyly. (b) Anteroposterior radiograph of the same hand.

36.2 Diagnosis

The diagnosis of polydactyly often will be evident on presentation. Investigations will differentiate the skeletal involvement of the polydactylous digit(s) based on both clinical examination and standard radiographs. Radiographs will determine the presence of an articulation of the polydactylous digit, the level of articulation, and the presence of an additional metacarpal (► Fig. 36.6). During the clinical examination, it is important to expose all limbs to identify additional polydactylies. Radial and ulnar polydactylies may be isolated or part of a syndrome; as such, a thorough examination of the child apart from the extremities is warranted to rule out other underlying



Fig. 36.3 Example of a distal type III polydactyly of the index finger.

malformations. Special attention for café au lait spots and short stature is warranted in radial polydactyly patients due to the association with Fanconi anemia.

36.3 Nonoperative Management

36.3.1 Radial Polydactyly

Radial polydactyly is rarely managed nonoperatively. In mild cases of a Wassel type I deformity with a bifid distal phalanx resulting only in a widened thumb tip and minimal nail abnormality, observation may be reasonable. Otherwise, surgical intervention is recommended for radial polydactyly.

36.3.2 Central Polydactyly

Surgical management is the mainstay for treatment of central polydactyly. However, if only a small soft-tissue mass is present, ligation may be an option. For more information, see the Ligation section for ulnar polydactyly.

36.3.3 Ulnar Polydactyly

Observation

Observation is an option for both types A and B ulnar polydactyly; however, the supernumerary digit can cause functional deficits, particularly with pedunculated digits that can catch on objects. Observation is likely most appropriate when a nubbin is present and is asymptomatic.

Ligation

Other nonsurgical options for type B include ligation, usually with a 4.0 silk suture or a surgical clip. Ligation has traditionally been the most common form of treatment for pedunculated type B digits, producing necrosis of the digit and subsequent autoamputation. Ligation can be done in the nursery shortly after delivery, but can be performed at a later stage ranging from 2 to 40 weeks (average 8 weeks of age). Rayan et al



Fig. 36.4 A clinical example of type A ulnar polydactyly.



Fig. 36.5 A clinical example of ulnar type B polydactyly.



Fig. 36.6 Anteroposterior radiograph of type A ulnar polydactyly.

recommended ligation only for digits with narrow stalks with suture placement as close to the finger to limit nubbin formation. Additionally, Mullick et al recommend limiting this treatment to stalks <2 mm with the presence of any bony or ligamentous attachments as a contraindication to ligation. Close observation for bleeding and infection and a discussion with parents about the risk of potential revision procedures are also important components of this management strategy. This technique has become controversial due to the concern of associated complications (see Complications section). A recent poll of pediatricians and hand surgeons in the United Kingdom on referral to a hand surgeon versus ligation in the neonatal period demonstrated that the majority of clinicians would refer for surgical assessment. However, proponents of ligation cite its low cost and simplicity.

36.4 Operative Treatment

36.4.1 Radial Polydactyly

Treatment of radial polydactyly is typically surgical and can be initiated at 6 to 9 months of age. The goals of surgery are both functional and cosmetic. Preoperative surgical splinting has been found to be valuable for improving angular and rotatory deformities prior to surgical correction. There are two primary surgical treatment options: (1) ablation with collateral ligament reconstruction or (2) the Bilhaut–Cloquet procedure.

Ablation with collateral ligament reconstruction is preferred for duplications with asymmetry resulting in the presence of a larger dominant thumb and smaller hypoplastic thumb. This most commonly includes removal of the radial (hypoplastic) duplicate, centralization of the flexor and extensor mechanism, collateral ligament reconstruction, and correctional osteotomies for angular deviation. Removing the radial duplicate when possible leaves the ulnar collateral ligament intact, which is of primary importance in thumb stability and pinch (► Fig. 36.7). In a type IV thumb, partial resection of the metacarpal head may be necessary for proper alignment of the phalanx on the metacarpal head. Therapy following surgery usually includes scar management and nighttime splinting for a period of 3 to 6 months. In cases where significant reconstruction of the radial collateral ligament is required, a full-time thumb spica splint may be worn for 2 to 3 months for additional stability. It should be mentioned ablation alone is not recommended for radial polydactyly treatment as it uniformly leads to joint instability.

The Bilhaut–Cloquet procedure was originally described in 1889. It involved the excision of the central portion of a type I thumb duplication with preservation of the ulnar aspect of the ulnar most portion and the radial aspect of the radial most portion of the duplicated distal phalanges. The idea was to allow a normal-size digit to be created from the two abnormal portions while maintaining the collateral ligaments for stability. It was initially used for type I and II deformities but expanded to types

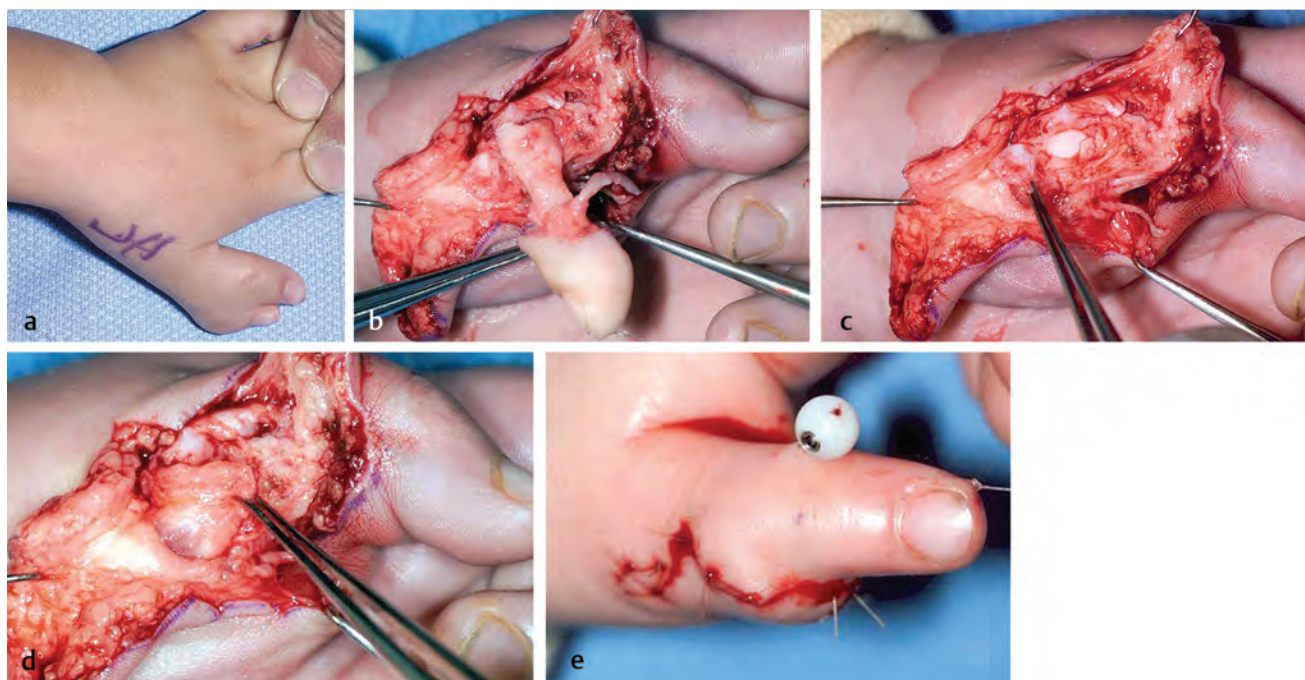


Fig. 36.7 (a) Surgical reconstruction of the collateral ligament in a 14-month-old child with Wassel type IV thumb duplication. (b) The radial duplicate is dissected free from the preserved ulnar thumb. (c) During dissection, the radial collateral ligament of the radial thumb, along with a piece of cartilage and periosteum, is preserved. (d) This flap of tissue is then imbricated into the radial aspect of the retained thumb. Sutures can often be passed through the bone and periosteum surrounding the base of the metacarpal. (e) The metacarpophalangeal joint is then immobilized for 3 to 4 weeks with a K-wire placed through the joint.



Fig. 36.8 Image of a 9-year-old girl treated with the Bilhaut-Cloquet technique for Wassel type II thumb. The reconstructed thumb shows residual nail grooving and minimal interphalangeal joint motion as evident by lack of skin wrinkling over joint.

III and IV. The Bilhaut-Cloquet procedure is technically difficult requiring exact alignment of the nail, bone, and growth plate (► Fig. 36.8). It may be recommended when the duplicated digits are similar in size. In an effort to avoid nail grooving and growth plate problems associated with the Bilhaut-Cloquet procedure, some surgeons recommend a modification in which the lateral components of the radial digit are maintained for digital width, but the entire nail is taken from the ulnar digit.

36.4.2 Central Polydactyly

Surgical care is usually initiated between 6 and 12 months of age (► Fig. 36.9). The goal is to begin surgical management before joint angulation and contractures occur. Despite an early start on surgical management, patients usually require multiple operations to obtain a cosmetically pleasing and functional hand. Because central polydactyly is so often seen in combination with syndactyly, radiographs are necessary to define the underlying bony anatomy and ideal surgical management.

In type II polydactyly, each phalanx has its own epiphysis. As the duplicated digit grows, it will displace the normal digit. Simple ablation is not appropriate and collateral ligament reconstruction is necessary. As the deformity worsens, corrective osteotomies and joint arthrodesis may be necessary. Tendonous attachments to both the digit to be maintained and that to be removed should be examined at the time of surgery. The most appropriate tendon should be retained. Accessory tendons may need to be transferred from the accessory digit to the retained proper digit.

36.4.3 Ulnar Polydactyly

The surgical treatment for both types A and B ulnar polydactyly ranges from soft-tissue excision to amputation of the polydactylous digit and may require augmentation of the soft tissues of retained digit. Functional outcomes are good, with minimal rehabilitation considerations.



Fig. 36.9 (a–d) Example of bilateral central polydactyly with corresponding anteroposterior (AP) radiographs. Surgical treatment of central polydactyly is complicated by differential growth of shared physis and potential for physeal injury during separation of the digits. (e) AP radiograph of both hands at the age of 7 years. (f–h) Functional and cosmetic result. Function is complicated by minimal motion at central digit proximal interphalangeal (PIP) and distal interphalangeal (DIP) joints, which has resulted in some scissoring of the digits.

Type A

Surgical ablation of the duplicated digit is indicated to improve hand function and aesthetics. An elliptical incision is planned around the digit. If a soft-tissue deficit is anticipated after ablation of the digit, a racquet-type incision can be made to use ulnar border skin of the polydactylous digit for augmenting soft-tissue coverage of the small finger. Incisions should be designed in a midaxial position when possible, as volar incisions have the potential to migrate and lead to flexion contractures. Exploration and division of the tendinous and neurovascular structures to the extra digit is critical while preserving the function and sensation to the remaining digit. Excision may also require soft-tissue release, tendon balancing, joint surface alignment, corrective osteotomies, and reattachment of intrinsic muscles.

Type B

Surgical excision of pedunculated digits can be performed in the operating room using either a combination of oral sedation with topical and local anesthetic or general anesthesia after the age of 3 months. The polydactylous digit is placed under tension and an elliptical incision is made at the base. The vessel and nerve are identified. The vessel is coagulated and the nerve is dissected and transected proximally under tension to avoid neuroma formation. The digit is then excised completely. Absorbable suture can be used to close the wound.

36.5 Complications

36.5.1 Radial Polydactyly

The most common complications following surgery are instability or stiffness at either the interphalangeal (IP) or



Fig. 36.10 Clinical image of a late zigzag deformity in a patient with previous Wassel type IV thumb duplication. Long-term instability of thumb metacarpophalangeal (MP) joint has resulted in subsequent instability at interphalangeal and MP joints.

metacarpophalangeal (MCP) joint, growth plate injury, and angular deformity. First webspace narrowing, nail deformities, and pulp deficiencies may also be seen.

Goldfarb et al reviewed the results of 31 thumbs treated for duplication. They found significantly decreased nail width (111% of the index nail on the operative side compared to 136% on the nonoperative side), while girth and length were similar between the operative and nonoperative sides. Twenty-two thumbs demonstrated IP joint angulation, with 12 thumbs having more than 5 degrees of angulation. IP joint angulation negatively correlated with visual analog scale score.

Stutz et al evaluated 43 thumbs with an average of 17 years after initial treatment for radial polydactyly. Eight patients underwent reoperation an average 8 years after the initial operation. Five patients underwent IP joint arthrodesis, while three had extrinsic tendon realignments for zigzag deformities after type IV thumb reconstruction (► Fig. 36.10).

36.5.2 Central Polydactyly

Early physeal closure, limited digital range of motion, and joint instability are frequent complications for type II central polydactylies. Webspace and joint contractures are also common in central polydactyly and synpolydactyly.

Wood reviewed the University of Iowa experience treating central polydactyly over 50 years. Fifteen percent of all polydactyly cases involved the ring (26) or long (8) fingers. Thirteen of 22 patients had bilateral hand involvement. A positive family history of similar hand anomalies was present in 55% of patients. Twenty of 22 patients had a complex syndactyly with a type II polydactyly. Sixteen of 22 patients underwent 91 procedures. The most common complications included webspace and flexion contractures. Fused, deviated, or nonfunctional digits represented more severe complications (► Fig. 36.9).

In 1982, Tada et al published a series of 12 patients with central polydactyly. In the series, all were type II and 10 of 12 hands had associated syndactyly. In Japan, maintaining a five-digit hand is of paramount importance even if the digits are nonfunctional. Therefore, in the majority of patients, the authors removed the polydactylous digit at the first surgery and focused on improving function during subsequent surgeries. Primary complications included joint deviations and contractures as well as rotatory deformities, believed to be secondary to intrinsic muscle atrophy. The authors conclude that creation of a five-

digit hand usually led to poor functional results and they believe a four-digit hand with removal of the hypoplastic ray should be the goal of reconstruction in central polydactyly.

36.5.3 Ulnar Polydactyly

Most commonly, complications occur after ligation of an ulnar polydactyly. These include (1) residual nubbin or incomplete amputation that may or may not contain neuroma, (2) gangrenous digit that does not slough and requires a secondary procedure for removal, and (3) infection. Watson and Hennrikus reported infection in 1 of 21 patients, but a residual bump in 43%; yet these did not require surgical intervention. Mullick et al reported on 10 cases of residual neuroma after ligation that required a revision procedure for symptomatic relief.

Complications of surgical excision in type A ulnar polydactyly include unsightly or painful scar, intrinsic tightness if transfer was necessary, prominence of the metacarpal head, and instability of the MCP joint. However, these complications are less common than those associated with ligation.

36.6 Conclusion

Radial polydactyly is a relatively rare condition. The different types of thumb duplication are most commonly described by the Wassel classification. The majority of cases are treated surgically anywhere from 6 to 24 months of age depending on thumb type and surgeon preference. The two main treatment options are ablation of the hypoplastic duplicate thumb with soft-tissue reconstruction and the Bilhaut–Cloquet procedure (or modification). The most common complications are joint stiffness or instability.

Central polydactyly is relatively rare. It is often associated with syndactyly. Radiographic imaging is key to successful surgical planning, which is the mainstay of management for central polydactyly. The overall goal is to create a functional and aesthetically pleasing hand.

Type A ulnar polydactyly is relatively uncommon, while type B is relatively prevalent among African American patients. Type A polydactyly is managed with surgical excision. Although controversy exists over the most appropriate treatment option for type B polydactyly, ligation is generally accepted in cases of uncomplicated, narrow stalks.

36.7 Key Points

- Radial polydactyly is most commonly classified by the Wassel classification, of which type IV is the most common.
- The two main surgical options for management of radial polydactyly are ablation with collateral ligament reconstruction are the Bilhaut–Cloquet procedure.
- Central polydactyly is usually associated with syndactyly and, therefore, radiographs are key to surgical success.
- Ulnar polydactyly is classified as type A or B, with the latter being more common, particularly among African American patients.
- Management of type A ulnar polydactyly is usually surgical, while type B is often managed surgically, with ligation an option in appropriate cases.

Suggested Readings

Goldfarb: A detailed technique article for treating radial polydactyly. The manuscript addresses indications, contraindications, and complications; 2006

Stutz, et al: A review of 43 surgically reconstructed thumbs > 10 years after treatment. Two scoring systems, reoperation rates, as well as pinch strength are reported; 2014

Mills, et al: A review of the outcomes and cost-effectiveness of the use of a surgical clip for type B ulnar polydactyly; 2014

Wood: A seminal paper on central polydactyly reviewing frequencies of each type of polydactyly as well as surgical management and outcomes; 1971

Rayan, et al: A retrospective review of a large series of ulnar polydactylous digits. Management and outcomes are discussed as well as an in-depth introduction to ulnar polydactyly pathology and epidemiology; 2001

Abzug, et al: A recent review of the evidence supporting management of type B ulnar polydactyly with a discussion on ligation of the polydactylous digit; 2013

Pritsch, et al: This article reviews and classifies type A ulnar polydactyly with a thorough introduction to the pathology; 2013

37 Macroductyly

Carolyn M. Pike and Brian I. Labow

Summary

Macroductyly is a rare congenital overgrowth disorder of the upper or lower limb. Although all tissue types are affected variably, proportion and patterning are maintained within the affected territory, and structures appear grossly normal. The disease process often follows a sensory nerve distribution within the hand or foot. Recent genetic studies have identified somatic mosaic gain-of-function mutations in the kinase *PIK3CA* in patients with macroductyly, as well as a number of related overgrowth conditions. However, the basis for phenotypic variation, such as severity and tissue distribution of overgrowth, as well as rate of growth, remains to be established. The diagnosis of macroductyly should be reserved for patients with a sporadic, isolated congenital digital enlargement affecting all tissue types not attributable to a syndrome or underlying vascular anomaly. Current treatment options are limited to surgery and can include staged debulking of soft tissue, epiphysiodesis, osteotomy, or ray amputation. Longitudinal growth is far easier to correct than circumferential growth. The major goals of treatment are to enable optimal hand function and to provide an aesthetic limb. Minor complications of operative treatment include hypertrophic scarring, decreased sensation, and flexion contractures. As patients age, many will develop progressive stiffness in the affected digit, and some will show radiological signs of a progressive osteoarthritis. In the future, pharmacotherapy may provide an additional treatment option for patients with digital overgrowth.

Keywords: macroductyly, overgrowth, limb, congenital

37.1 Introduction

Macroductyly is a rare congenital overgrowth disorder affecting the digits of the upper or lower extremity. The condition accounts for 0.9% of all upper limb congenital anomalies. Macroductyly is sporadic, nonsyndromic, and often follows a sensory nerve distribution within the hand or foot. Overgrowth is usually noted at birth; however, both the extent and rate of growth can vary widely. Macroductyly can be classified by rate of growth, with the term “progressive” describing rapid, disproportionate enlargement relative to the rest of the limb, while “static” refers to a slower, proportionate growth. Although all tissue types are affected variably, proportion and patterning are maintained within the affected territory, and structures appear grossly normal. Another means of classification has been through the predominant tissue type involved, for example, lipomatous macroductyly for cases with gross excess of adipose tissue out of proportion to other tissue types. In contrast, nerve-territory-oriented macroductyly has been used for digits with enlarged nerves within the affected zone of overgrowth that clearly follow a sensory nerve territory. A biological distinction between these two clinical entities has not been shown. To further confound the situation, a wide array of descriptive terms and eponyms has also been used to refer to macroductyly. Given

the rarity of this condition, the lack of precision in nomenclature, and our limited understanding of the pathophysiology of this disorder, it is not surprising that there exists a paucity of literature on treatment options and long-term outcomes.

37.2 Diagnosis

In most cases, digital enlargement is obvious soon after birth. Occasionally, minor degrees of enlargement may not be detected until later in infancy, when continued enlargement occurs (► Fig. 37.1, ► Fig. 37.2). A variety of syndromic and non-syndromic causes for digital enlargement exist. A thorough physical examination to look for other findings or a local or regional cause for the overgrowth (e.g., expansile mass, vascular anomaly) should be carefully undertaken. The diagnosis of macroductyly should be reserved for patients with a sporadic, isolated congenital digital enlargement affecting all tissue types not attributable to a syndrome or underlying vascular anomaly. Radiographic imaging such as ultrasonography or plain films can be useful to document the absence of a vascular lesion and the presence of skeletal involvement, respectively. Magnetic resonance imaging can also be used to plan debulking procedures by documenting the extent of fatty overgrowth in the limb that frequently contributes to additional limb or digit girth and weight. Recent genetic studies have identified somatic mosaic gain-of-function mutations in the kinase *PIK3CA* in patients with macroductyly. These mutations have been implicated in a growing number of related overgrowth conditions as



Fig. 37.1 A 13-month-old male with severe macroductyly and syndactyly of the index and middle fingers. The overgrowth includes the thumb and adjacent palm and follows a roughly median nerve sensory pattern.

well. However, the basis for phenotypic variation, such as severity and tissue distribution of overgrowth, as well as rate of growth, remains to be established.

37.3 Nonoperative Treatment

Since macrodactyly typically gives rise to a poorly functioning and aesthetically displeasing digit, treatment is rarely nonoperative. If the affected digit is mildly overgrown with adequate function and is pain free, then observation should be recommended. In some cases, therapy may help with digits that are mildly stiff, though progressive arthritic changes may still occur.



Fig. 37.2 A 15-month-old female with mild macrodactyly affecting the index finger. The overgrowth is predominately circumferential with minimal length difference as compared to the contralateral side.

In the future, it is possible that pharmacological inhibitors of the PI3K-AKT pathway may halt disease progression or prevent recurrence in postoperative patients. Small clinical trials have been initiated with drugs such as rapamycin, but there have been limited results published. For the most part, current treatment options remain limited to surgery.

37.4 Operative Treatment

Surgical treatment for patients with macrodactyly should be individualized. The patient's age, digit involved, type and rate of overgrowth, as well as parental and surgeon preferences will all be factors in determining the treatment plan. The major goals of treatment are to enable optimal hand function and to provide an aesthetic limb. In some instances, that may mean amputation. However, a variety of procedures have been described that reduce the length and circumferential bulk of the digit, while maintaining sensation and circulation within the extremity.

An overgrown digit may be salvaged via skeletal or soft-tissue reduction, or a combination of the two. In some instances, several procedures may be performed over time. Longitudinal excess is far easier to correct than circumferential excess, and it should be noted that these digits never look or function normally (► Fig. 37.3). Adjunct procedures such as epiphyseal growth arrest and osteotomies can also be performed to correct angular deformities. Debulking procedures should be limited to one side of a digit at a time to minimize the risk of vascular compromise and usually require a staged approach.

If the digit is characterized as progressive macrodactyly with a grossly overgrown appearance and minimal functionality that often interferes with the other digits, amputation may be considered. Generally, a complete ray resection or transmetacarpal amputation is required. If the thumb must be amputated to the level of the metacarpal, a minimally affected index ray can be pollicized or a microvascular toe transfer can be considered.

37.5 Complications

Minor complications of operative treatment include hypertrophic scarring, decreased sensation, and flexion contractures. After repeated debulking and nerve manipulation, protective



Fig. 37.3 (a) A 7-year-old female with ulnar-sided macrodactyly affecting the small and ring fingers predominantly. (b) One year following soft-tissue debulking and closing wedge osteotomy of the small finger. The ring finger treated at another institution was managed with partial middle and distal phalangectomies and distal interphalangeal joint fusion. Although the length discrepancy has been improved, circumferential overgrowth remains marked.

sensation is typically the result. As patients age, many will develop progressive stiffness in the affected digit and some will show radiological signs of a progressive osteoarthritis. Early arthritis in toes transferred to the hand has also been observed.

37.6 Conclusion

The rarity of macrodactyly and the highly variable phenotypic presentation have hindered both descriptive and comparative studies on treatment options and long-term outcomes. In general, treatment for macrodactyly should be viewed as palliative, although surgical therapy can certainly benefit many patients. Depending on clinical circumstances and patient or parent preference, debulking techniques and amputation both have a role in managing the affected digits. Although reconstruction will not result in a fully normal digit and often require multiple operations, they are well-tolerated procedures and may offer a restoration of function and aesthetics. In the near future, pharmacological options may exist to abrogate digital overgrowth or prevent additional growth after surgical debulking.

37.7 Key Points

- Macrodactyly is rare and characterized by congenital distal overgrowth of the upper or lower limb. All tissue types are affected to a variable degree.
- Recent studies have identified somatic mosaic mutations in the *PIK3CA* gene in patients with macrodactyly.
- Current treatment options are limited to surgery and can include staged debulking of soft tissues, epiphysiodesis, osteotomy, or ray amputation.
- In general, treatment options are well tolerated with few minor complications; however, patients and families should be counseled that no treatment is entirely satisfactory.

Suggested Readings

Cerrato F, Eberlin KR, Waters P, Upton J, Taghinia A, Labow BI. Presentation and treatment of macrodactyly in children. *J Hand Surg Am.* 2013; 38(11):2112–2123

Carty MJ, Taghinia A, Upton J. Overgrowth conditions: a diagnostic and therapeutic conundrum. *Hand Clin.* 2009; 25(2):229–245

38 Brachial Plexus Palsies

Raymond Tse and Angelo B. Lipira

Summary

Pediatric brachial plexus palsies can present at birth (1 in 1000 newborns) and from blunt trauma in older children. The extent of involvement and severity of injury is variable. This chapter will discuss assessment, nonsurgical treatment, and surgical treatment of brachial plexus palsy.

The approach to surgical exploration will be detailed and a spectrum of scenarios will be presented so that the principles of primary nerve reconstruction (including nerve graft and nerve transfers) can be illustrated. These include upper plexus, pan-plexus, multiple root avulsions, isolated deficits, delayed presentation and failed reconstruction. Technical details of nerve grafting and nerve transfers will be described.

Secondary musculoskeletal consequences of brachial plexus palsy will also be discussed including strategies for prevention and options for secondary surgical reconstruction.

Keywords: brachial plexus palsy, Erb palsy, glenohumeral dysplasia, nerve graft, nerve transfer

38.1 Introduction

The brachial plexus is a complex network of nerves that provide sensation and motor control of the upper extremity. In addition to the intricacies of its form and functions, growth and development in the setting of chronic denervation add another layer of complexity to the management of pediatric brachial plexus palsies.

An understanding of the anatomy and presentations is important in the evaluation of patients. An appreciation of the natural history and outcomes is critical when deciding upon treatment. Preservation of musculoskeletal health in the setting of chronic denervation is needed to optimize functional results.

Given the many dimensions of pediatric brachial plexus palsy, children are best served through a multidisciplinary team approach at specialized centers offering coordinated, longitudinal care. Early referral allows for better clinical assessment and may reduce the chances of sequelae that affect long-term outcomes.

38.2 Presentation

Neonatal brachial plexus palsy (NBPP) occurs in 1/1,000 live births and presents immediately after delivery. Downward traction on the brachial plexus results in injuries of the upper plexus. With further traction, the zone of injury progresses to lower regions of the plexus, ultimately resulting in pan-plexus palsy in its most severe expression. Presentations can generally be grouped into four patterns: type I involves C5 and C6 deficits (Erb/Duchenne type) with loss of shoulder abduction, shoulder external rotation, elbow flexion, and forearm supination. The limb assumes the Erb posture due to intact antagonist muscles. Type II involves C5–C7/C8 deficits (extended Erb type), resulting in the additional loss of wrist extension. The limb assumes the

“waiter’s tip” posture. Type III involves deficits from C5 to C8/T1, resulting in an arm that is generally flaccid or paralyzed. Type IV involves C5 to T1 as well as the sympathetic chain, resulting in flail arm with a Horner syndrome.

Upward traction on the brachial plexus results in injuries of the lower plexus, resulting in loss of hand and wrist function with intact elbow and shoulder function. This presentation is known as Klumpke’s palsy and is extremely rare in NBPP.

When a root level is affected, the site of injury tends to be at the trunk level in the upper plexus and more proximal in the lower plexus. This may be related to the presence of fascial sleeves that extend from the bony vertebrae to the extraforaminal nerve roots in the upper plexus. This fascia is progressively absent at more inferior nerve root levels. As such, lesions of the lower roots are more likely to be preganglionic spinal root avulsions. One exception to this pattern occurs in breech deliveries, in which C5 and C6 lesions tend to be avulsions.

Traumatic brachial plexus palsy (TBPP) can occur in older children, but is rare. The mechanism is usually blunt injury and most often results from motorized vehicle accidents. Compared to NBPP, the forces involved are rapid and high energy. Consequently, the patterns of injury in TBPP are varied and more likely involve spinal root avulsions.

Injuries associated with NBPP can include fractures of clavicle, humerus, and rarely cervical spine. Phrenic nerve injury can occur with severe palsies, and hypoxic brain injury can result from asphyxia. Children with TBPP should undergo a general trauma survey.

Several conditions may present with altered limb posture or movement and must be differentiated from brachial plexus palsy. “Pseudoparalysis” can result from pain related to bony fracture; however, once healed, normal limb function resumes. A septic shoulder may present with loss of shoulder movement, but fever, constitutional symptoms, and a “hot joint” will often be present. Congenital limb differences such as arthrogryposis can be distinguished by skeletal hypoplasia, absent skin creases across joints, and associated anomalies. Lesions of the central nervous system may present with altered tone, lower extremity findings, global cognitive impairment, and/or progressive weakness.

38.3 Assessment

38.3.1 Initial Assessment

Initial history focuses on birth, associated injuries, limb presentation, and progression of recovery. Specific limb postures should be elicited, given many families will describe a flaccid arm in spite of intact motor functions. For example, when hand function is present, it can be helpful to understand if the hand function has always been present (i.e., a Type 1 palsy) or if that function has recovered (i.e., a Type 3 palsy with persistent upper plexus deficits). Any history of recovery helps understand the type of nerve injury that has occurred. Neuropraxia (Sunderland 1) involves no Wallerian degeneration, and function recovers rapidly. Axonotmesis (Sunderland 2) involves

Wallerian degeneration and requires axon regrowth. In general, recovery of full active range of motion within 1 month of age is associated with no measurable long-term deficits in strength, sensation, or limb growth. Slower recovery (i.e., Sunderland 3 and 4 injuries) is associated with greater long-term deficits. Complete nerve disruption (Sunderland 5) results in no recovery.

Initial examination includes an assessment of global cortical function including age-appropriate developmental milestones, trunk control, tone, and posturing. Neurologic examination of lower extremities helps rule out central nervous system lesions. Respiratory function should be assessed to screen for associated phrenic nerve injury. The face should be assessed for Horner syndrome (ptosis, myosis, anhydrosis) that results from disruption of the sympathetic chain. Given that the most cephalic contribution to the sympathetic chain exits the spinal cord along the T1 foramen, persistent Horner syndrome associated with pan plexus palsy suggests a proximal T1 injury and possibly an avulsion. Scapular stability and rhomboid function should be assessed to determine involvement of dorsal scapular nerve. Given the very proximal divergence of this nerve from the brachial plexus, a deficit suggests a proximal zone of injury that may involve a preganglionic root avulsion. Infants with NBPP should also be assessed for torticollis, which is frequently present and may be related to traction injury to neck musculature.

Focused examination of the brachial plexus follows the more general initial exam. Examination of infants with NBPP is limited to observed and encouraged movements and these can be rated according to the Active Movement Scale (AMS; ► Table 38.1). Movement at each major limb articulation is examined and recorded at each visit. Infants may or may not “perform” on a given day and repeated examination helps in developing a more reliable assessment.

Older cooperative children with TBPP may undergo more detailed examination including sensation and specific muscle group testing. Both AMS and British Medical Research Council (MRC) scale can be used to record motor function. The Mallet scale measures composite motions that are important for activities of daily life, and is useful for assessing outcomes and for determining the need for secondary procedures in children with both NBPP and TBPP (► Fig. 38.1).

One of the difficulties in management of nerve injuries is the uncertainty of a single examination. The spectrum of the extent

of injury (i.e., zone of injury), severity (i.e., Sunderland class), and the fact that variations in injury severity can exist in different regions of a patient’s brachial plexus make the prediction of outcome complex. In infants with NBPP, electromyogram (EMG) has been found to be “overly optimistic.” As such, an appreciation of the severity of nerve injury relies upon repeated longitudinal assessments.

Early assessment within 1 month of age or injury establishes a baseline, and repeated monthly or bimonthly examination helps understand the pattern of recovery. Assessment by a consistent examiner helps reduce variation. Ongoing therapy and nonsurgical treatment are critical and should be incorporated into assessments and longitudinal care.

38.4 Nonsurgical Treatment, Natural History, and Indications for Nerve Surgery

38.4.1 Nonsurgical Treatment

All children with brachial plexus palsy should receive nonsurgical treatment with therapy to maintain passive range of motion, encourage limb awareness, maintain sensory feedback, and facilitate cortical development and functional adaptation. Dedicated physical and occupational therapists, as well as rehabilitation specialists with expertise in pediatric brachial plexus palsy are central in these efforts. Nonsurgical treatment may be more important to outcomes than surgical treatment, given that contractures and limb neglect can negate any improvements from nerve reconstruction. More detailed discussions of strategies to avoid musculoskeletal complications follow (see section “Musculoskeletal considerations and complications”).

The decision to pursue nerve reconstruction is based on the fundamental question of whether the results with surgery will be better than nonsurgical treatment alone. Given that nerve reconstruction is imperfect and final outcomes are uncertain, an understanding of natural history and surgical outcomes is important in deciding a course of treatment.

38.4.2 Indications for Surgical Exploration: Neonatal Brachial Plexus Palsy

The majority (70–90%) of infants with NBPP rapidly recover adequate function that nerve exploration is not indicated.

Most agree that a flail arm (i.e., flaccid limb with no hand function) associated with a persistent Horner syndrome beyond 1 month of age portends to a poor prognosis. The association of Horner syndrome with no hand function suggests a proximal injury of T1 (i.e., nerve root avulsion). There is a consensus that in this situation, early exploration and reconstruction are warranted in attempt to achieve reinnervation of hand functions.

Tassin (1983) studied a cohort of children with NBPP and found that infants who did not recover any elbow flexion by 3 months tended to have poor long-term shoulder and elbow function. Based on that study, Gilbert recommended surgical exploration for any infant who did not recover biceps by 3 months of age. Although Gilbert’s reported outcomes are favorable, these have not been compared to outcomes without surgery.

Table 38.1 Active movement scale

Score	Gravity	Observation
0	Gravity eliminated	No contraction
1	Gravity eliminated	Contraction, no motion
2	Gravity eliminated	≤ 1/2 active range of motion
3	Gravity eliminated	> 1/2 active range of motion
4	Gravity eliminated	Full active range of motion
5	Against gravity	≤ 1/2 active range of motion
6	Against gravity	> 1/2 active range of motion
7	Against gravity	Full active range of motion

Mallet score legend

Abduction

External rotation

Hand to head

Hand to back

Hand to mouth

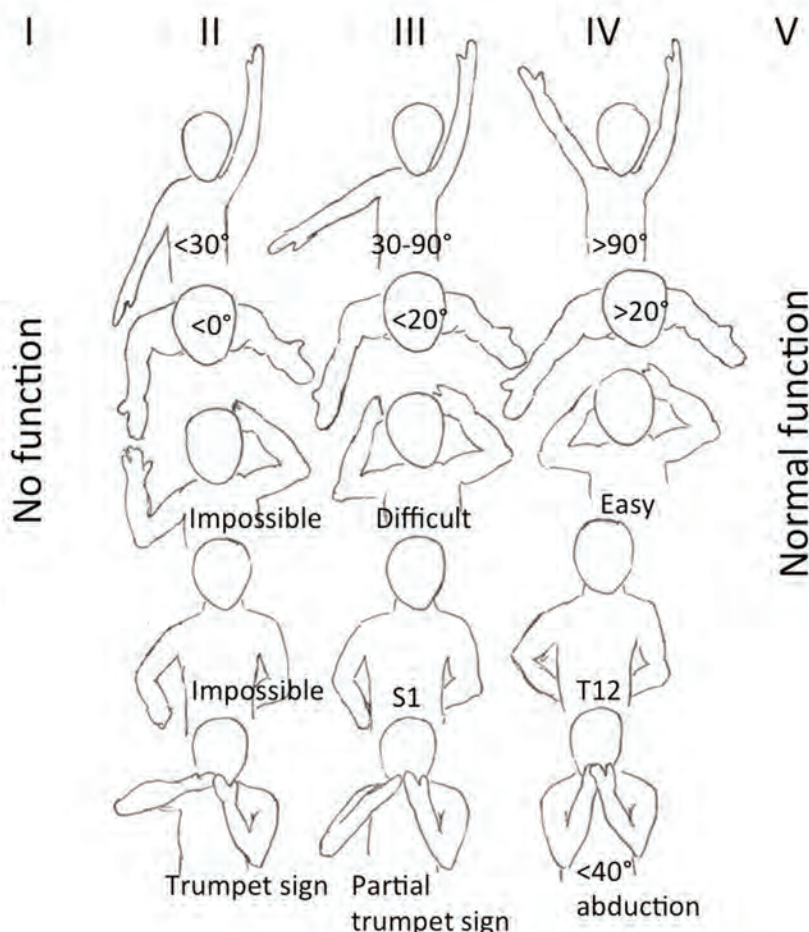


Fig. 38.1 Mallet scale.

Both Carter (2004) and Clarke (2007) have suggested that absence of elbow flexion at 3 months may be an overly aggressive indication for surgical treatment. Waters (1999) compared the results of microsurgical reconstruction with outcomes of nonsurgical treatment and stratified patients by the month after which elbow flexion recovered. He found that when elbow flexion takes greater than 5 months to recover, outcomes are better with surgery. He recommends the absence of elbow flexion within the first 6 months of life as an indication for nerve reconstruction.

Surgical indications based on the absence of elbow flexion or “biceps function” simplify recovery of the brachial plexus to a single clinical finding. Whereas simplifying decision-making for a complex condition has merit, our preferred approach considers all of the movements of the limb.

Clarke (1994) examined a cohort of patients with NBPP treated nonsurgically and analyzed all limb active movements at 3 months of age using the Toronto scale. Although lack of elbow flexion was the most significant predictor of a poor outcome, the lack of wrist, finger, thumb, and elbow extensions

were also independent predictors of poor outcome. These findings led to a statistical model in which a composite AMS score for these movements (elbow flexion, elbow extension, wrist extension, thumb extension, and finger extension) of less than 3.5/10 always predicted a poor outcome, and was thus an indication for surgical treatment. The clinical picture of this score is that of C5–C8 or worse deficits (i.e., pan-plexus or near pan-plexus palsy) at 3 months of age. These indications did not account for all patients with a poor outcome. As such, an additional test at 9 months was added for those infants who had adequate scores at 3 months of age. If a child was unable to pass the “cookie test” (the composite movement of getting hand to mouth with arm in an adducted position), they were offered surgical treatment. Fifteen years later, Clarke published results of nerve reconstruction (2009) suggesting that the results of nerve grafting were better than neurolysis (and presumably no treatment) based on the above indications for surgery. The Toronto protocol has evolved to include the lack of elbow flexion at 6 months as another indication for surgical treatment (► Table 38.2).

Table 38.2 Indications for nerve exploration and reconstruction (the Toronto protocol)

Age	Findings
1 mo	Flail arm with Horner syndrome
3 mo	Composite AMS score for elbow flexion, elbow extension, wrist extension, finger extension, and thumb extension of <3.5/10
6 mo	No elbow flexion
9 mo	Failed “cookie test”

Abbreviation: AMS, active movement scale.

38.4.3 Indications for Surgical Exploration: Traumatic Brachial Plexus Palsy

Older children with TBPP need to be considered on an individual basis given that there is little data to guide generalized treatment. The rapid, high-energy nature of TBPP results in greater variations of presentation and a greater likelihood of root avulsions. Older children have less capacity for nerve regeneration and the distance for axons to reach target is greater. In addition, development is not as rapid and cortical plasticity may not be as great. Principles applied to adult brachial plexus palsy may be adapted to the management of children with TBPP.

The presence of a Tinel's sign and careful clinical examination can help localize the specific site of injury. Given that axons grow at a rate of 1 mm per day, the measured distance from site of injury to target muscle can be used as a guide to determine how long an axonotmetic injury would be expected to recover. Exploration is considered if there is no evidence of nerve recovery beyond this time. As opposed to infants, serial EMG provides insights into potential recovery and should be used as an adjunct to clinical exam.

38.4.4 Investigations Prior to Nerve Exploration and Reconstruction

Once the decision to proceed with surgery has been made, assessment for nerve root avulsion can help with surgical planning. Nerve root avulsions are preganglionic lesions in which discontinuity of neurons from the spinal cord renders the root nonfunctional and without the possibility of functional end target reinnervation. A pseudomeningocele with absent rootlets is highly predictive of an avulsion. Computed tomography (CT) myelogram has been the standard imaging modality; however, this requires a lumbar puncture and significant radiation exposure in a young infant. We have compared newer techniques of magnetic resonance (MR) myelography and have found them to be equal in accuracy to CT myelogram (Tse 2014). MR myelography does not require lumbar puncture and avoids radiation exposure. Another advantage of MR is that bilateral shoulders can be included in the study to assess for glenohumeral dysplasia (GHD), skeletal changes that occur with chronic muscular imbalance. We have examined scans of infant shoulders and have found a very high incidence of skeletal changes (74%) that can occur early and in the absence of limited passive range of

motion. If significant skeletal changes are detected, the method of postoperative immobilization can be modified to address these abnormalities.

If nerve transfers are anticipated, EMG can be used to confirm suitability of potential donor nerves for transfer, given that some donor nerves can be affected by the brachial plexus palsy. For example, involvement of C7 results in partial denervation of the triceps muscles and can affect results of radial to axillary nerve transfers. EMG is used to assess the status of potential donors and to select the best one.

Ultrasound of the diaphragm is routinely performed to assess the functional status of the phrenic nerve, which can be intertwined with injured nerve elements and result in limited respiratory reserve.

38.5 Nerve Exploration and Reconstruction

38.5.1 Positioning

The posterior triangle of the neck can be accessed in the supine position by turning the head to the opposite shoulder (neck rotation and flexion; ► Fig. 38.2). A midline bump supports the kyphotic curvature of the lower cervical and upper thoracic spine to bring the plane of the brachial plexus parallel to the floor. The arm, shoulder, neck, and chest on the affected side should be exposed. Dissection is carried out with shoulder adducted to maximize the costoclavicular space.

38.5.2 Exposure

Supraclavicular Region

A horizontal incision is designed along a skin crease parallel to the clavicle, extending from the clavicular insertion of the sternocleidomastoid to the medial insertion of the trapezius. A vertical limb parallels the posterior border of the sternocleidomastoid and extends approximately midway to the mastoid (► Fig. 38.2). Although a single horizontal incision can be used, the exposure is more limited and relies upon soft-tissue retraction.

**Fig. 38.2** Brachial plexus exploration: positioning and incision.

The initial incision is made through platysma, and the superolaterally based flap is elevated in a subplatysmal plane. The platysma is thin and difficult to visualize in infants, but can be differentiated from overlying subcutaneous fat and underlying supraclavicular fat pad by its white smooth texture (► Fig. 38.3). The external jugular vein and the cervical plexus of sensory nerves are deep to the platysma and can also help define the appropriate plane of dissection. The external jugular vein needs to be retracted or ligated and the cervical plexus should be lysed from the network of veins intertwined with it so that it can be later used for transfer or as graft material (it is

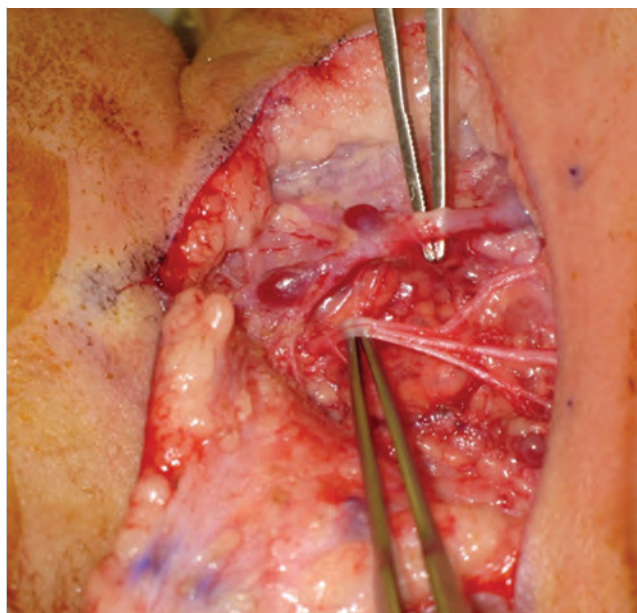


Fig. 38.3 Brachial plexus exploration: (the same patient and orientation as in ► Fig. 38.2) subplatysmal dissection to expose external jugular vein and cervical plexus. Note the shiny white appearance of the undersurface of platysma.

easier to clean the nerves before dividing distally). Each of the cervical plexus branches is traced proximally where they converge with each other and with the great auricular nerve (which courses over the anterior surface of sternocleidomastoid; ► Fig. 38.4a). Further dissection proximal reveals the C4 root and the phrenic nerve in a region outside of the zone of injury and severe scar (► Fig. 38.4b).

The C4 contribution to phrenic nerve is usually the most medial branch and can be found by gentle blunt dissection over the anterior surface of the anterior scalene. It takes a direct caudal course, paralleling the fibers of anterior scalene, and is under the prevertebral fascia. Its identity can be confirmed by electrical stimulation.

Once the phrenic nerve has been visualized, the supraclavicular fat pad is divided along the posterior border of the sternocleidomastoid and superior border of the clavicle. Bipolar cautery is used to reduce the possibility of lymphatic leak. Traction on the fat pad should be avoided, given this layer is continuous with the tissues that envelop the carotid sheath (whose contents can become unnecessarily exposed). Care should also be taken deep to the junction of sternocleidomastoid muscle and clavicle to avoid inadvertent injury to the thoracic duct or exposure of the subclavian vein. The bluish hue of the latter can sometimes be seen within the fat if the external jugular vein is followed too far caudal along its course. Clean incisions and preservation of the supraclavicular fat pad facilitates later repair, which is important in providing a protective layer for nerve reconstruction, an ideal gliding surface for nerve excursion, and a vascular environment for nerve grafts.

Deep to the fat pad, the transverse cervical artery and vein can usually be found running parallel to the clavicle. These vessels are often ligated for exposure. The omohyoid muscle is encountered at its tendinous midportion before exposing the brachial plexus. This muscle can be retracted or transected, and should be mobilized adequately to allow transposition so that it lies superficial to the supraclavicular fat pad at closure. This avoids muscle movements directly over newly reconstructed nerves.

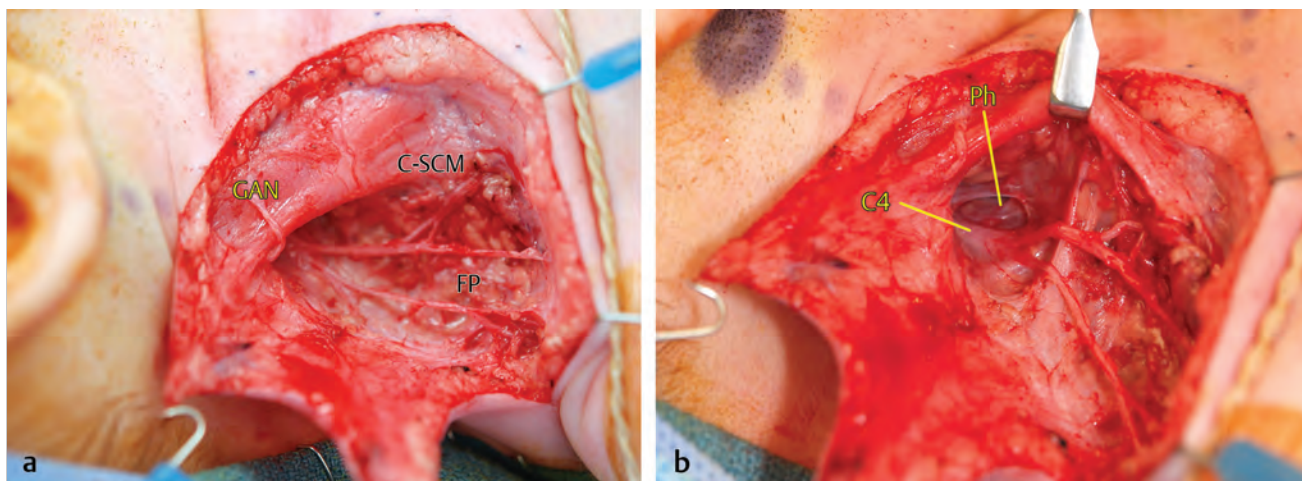


Fig. 38.4 Brachial plexus exploration. (a) The cervical plexus has lysed from the supraclavicular fat pad (FP). The great auricular nerve (asterisk) converges with cervical plexus branches proximally. The clavicular head of sternocleidomastoid (C-SCM) can be released for better inferior exposure if needed. (b) The cervical plexus (ph) can be traced proximally to the C4 root where the phrenic nerve can be identified in a region outside the zone of injury.

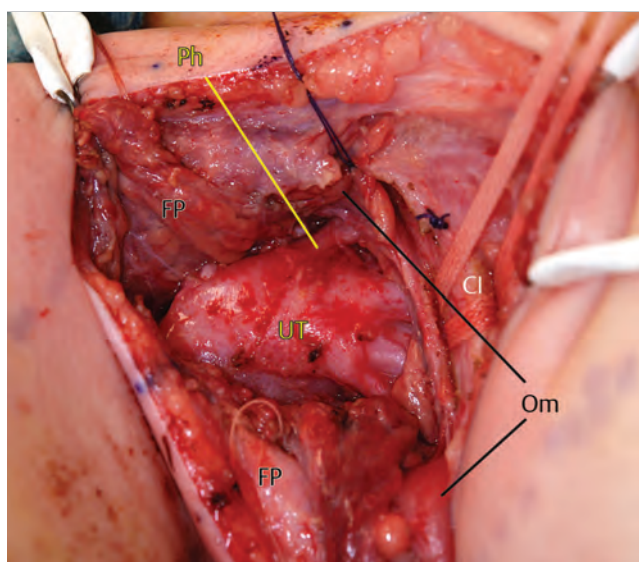


Fig. 38.5 Brachial plexus exploration. The omohyoid (Om) has been divided at its tendinous portion. The supraclavicular fat pad (FP) has been incised and elevated off of the brachial plexus. The clavicle (Cl) is being elevated by an umbilical tape to open the retroclavicular space. The phrenic nerve (Ph) is encased in the scar of the neuroma of upper trunk (UT).

The upper trunk of the brachial plexus is found just deep to the omohyoid muscle, and is often heavily scarred and distorted (► Fig. 38.5). Following the principle of working from “known to unknown,” the brachial plexus is exposed and defined starting outside of the zone of injury to facilitate the more difficult dissection through the scarred region.

The previously identified phrenic nerve can be followed along its caudal course from the C4 root on the anterior surface of the anterior scalene. As the nerve approaches the lateral border of muscle, it gives off a small branch to and receives a small branch from the C5 root. This interconnection is usually scarred and in most cases, a significant length of the phrenic nerve is enveloped in scar from upper trunk neuroma (► Fig. 38.5). Exposure between sternocleidomastoid and anterior scalene allows visualization of the distal course of phrenic nerve so that the phrenic nerve can be lysed from the neuroma. A thin layer of scar is left on the phrenic nerve to facilitate its safe handling. The interconnection between the phrenic nerve and the C5 root can be sacrificed without morbidity. With the phrenic nerve mobilized, the C5 and C6 nerve roots can be exposed. Partial release of the anterior scalene provides proximal exposure up to the bony intervertebral foramina. Dissection here should progress carefully given the rich venous plexus of the vertebral vessels just proximal. Care should also be taken to avoid traction on the phrenic nerve as it becomes particularly vulnerable with medial exposure.

Working from distal, the branches of the upper trunk can be identified under the clavicle where they are usually unscarred (► Fig. 38.6). The suprascapular nerve (SSN) parallels the omohyoid on its course toward the suprascapular notch and defines the lateral border of dissection. The anterior division of upper trunk (AD-UT) is the most medial branch and defines the medial border of dissection. The posterior division of upper

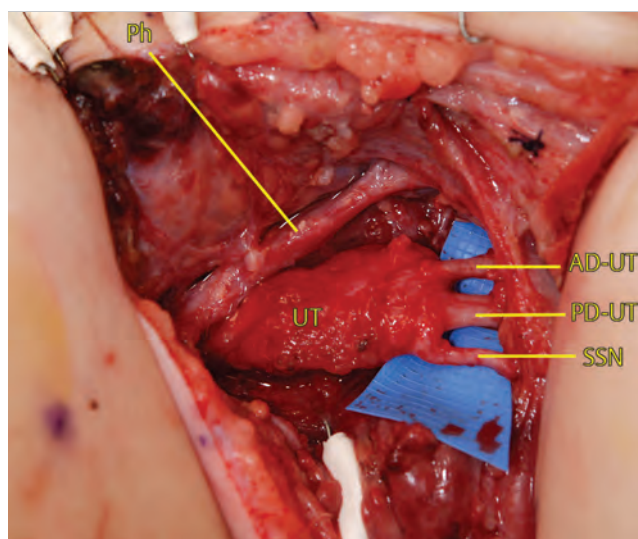


Fig. 38.6 Brachial plexus exploration: The phrenic nerve (Ph) has been lysed from the neuroma of upper trunk (UT). The distal branches, suprascapular nerve (SSN), posterior division of upper trunk (PD-UT), and anterior division of upper trunk (AD-UT) have been exposed and neurolysed.

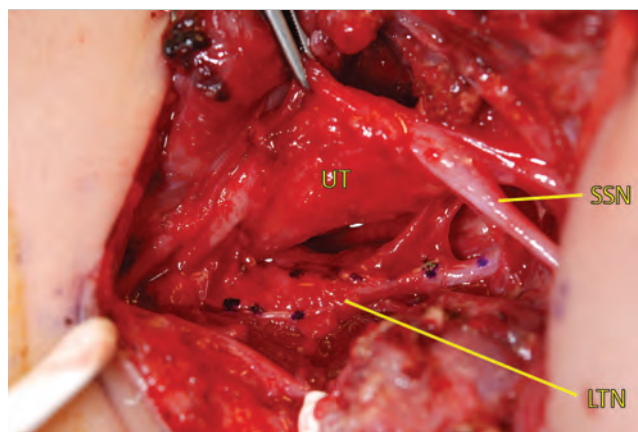


Fig. 38.7 Brachial plexus exploration. Branches from proximal nerve roots dive through the middle scalene to contribute to the long thoracic nerve (LTN). In this example image, some of the middle scalene muscle was inadvertently elevated with dissection on the posterior surface of the neuroma of upper trunk (UT) due to heavy scarification. Preservation of LTN is important to maintain scapular stability.

trunk (PD-UT) is found in the middle. The three branches can be followed to their convergence at the trifurcation of the upper trunk, which is usually found at the level of the clavicle.

The posterior surface of neuroma can be difficult to dissect free from the middle scalene muscle due to scarification. As the dissection progresses proximally, the planes become clearer; however, the C5, C6, and C7 contributions to the long thoracic nerve become vulnerable as these branches dive through the middle scalene muscle proximally (► Fig. 38.7). Preservation of these branches can often be achieved to avoid scapular instability.

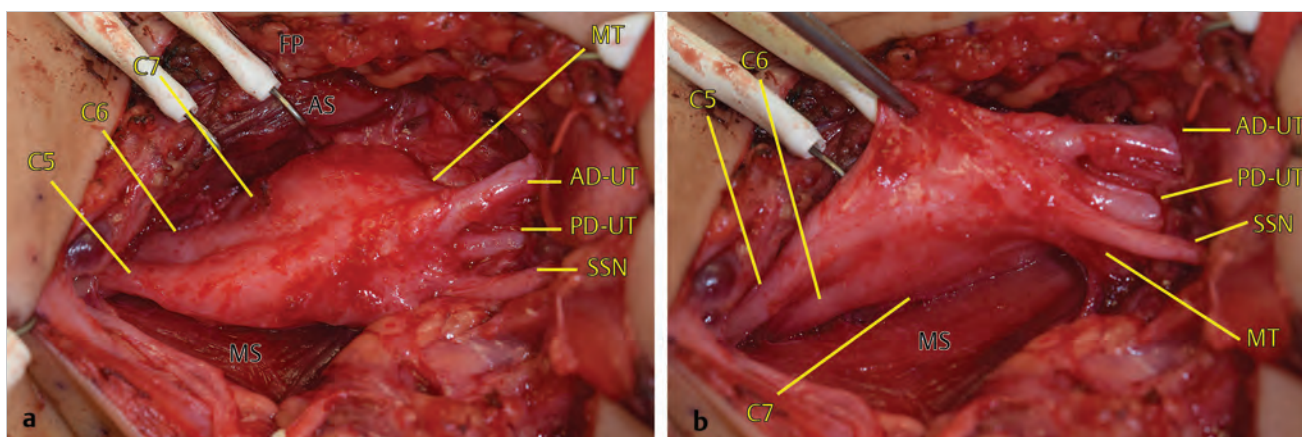


Fig. 38.8 Brachial plexus exploration. C7 can be found in the interval between anterior scalene (AS) and middle scalene (MS). In this case, there is a neuroma of the middle trunk (MT) that appears as a single scarred mass that includes the upper trunk (UT).

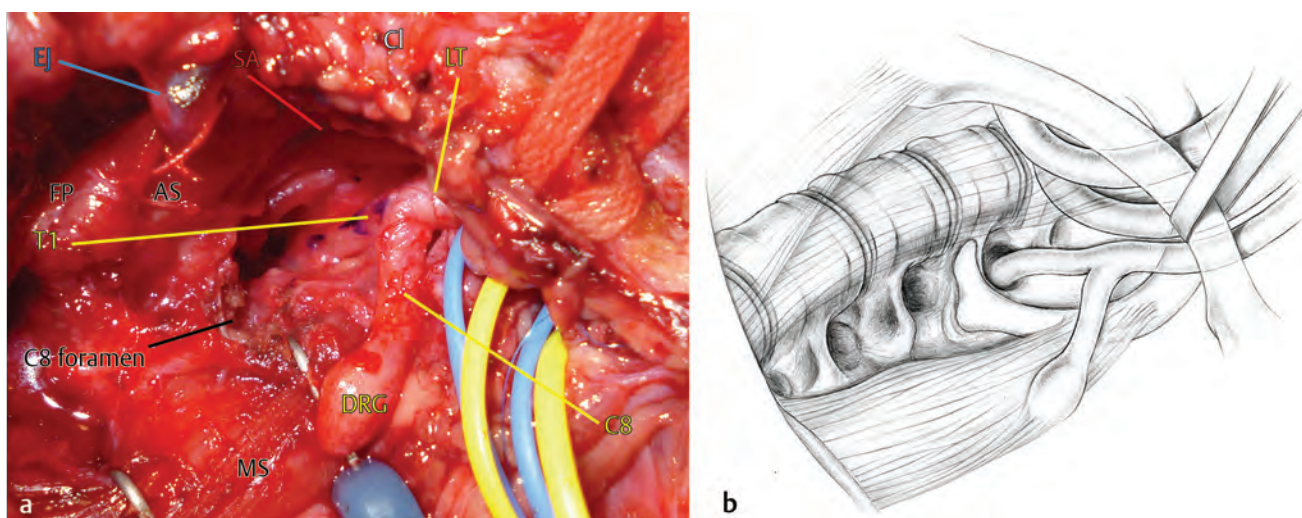


Fig. 38.9 Brachial plexus exploration. The lower roots are further caudal and, from the surgeon's perspective, deep in the retroclavicular exposure. In this case example, C8 has been avulsed and a dorsal root ganglion (DRG) is found outside of the C8 intervertebral foramen. The T1 root is marked with purple dots. The subclavian artery (SA) needs to be lysed from the brachial plexus. The lower external jugular (EJ) stump serves as a landmark for the subclavian vein, which courses anterior to the subclavian artery.

Retroclavicular Region

Due to the curvature of the spine, each sequential lower root is found inferior and dorsal to the root above. From the surgeon's perspective, each sequential lower root will seem to be deeper in the exposure. The middle trunk is identified behind the upper trunk (► Fig. 38.8).

Further deep and slightly inferior dissection reveals the lower trunk. Proximal dissection to and between the anterior and middle scalenes reveals the respective proximal nerve roots. The C8 and T1 roots can be traced proximally and can be seen originating above and below the first cervical rib, respectively (► Fig. 38.9). Dissection too far inferior and anterior will quickly lead to the subclavian artery. This structure can be easily mistaken for the lower trunk, but it is pulsatile and can be confirmed with Doppler if needed. The subclavian artery should be carefully lysed from the brachial plexus. The trunks of the brachial plexus normally divide into divisions at or beneath the clavicle. The divisions are rarely scarred or injured.

Although the vertebral artery is generally not visualized, it traverses ventral to the C7 root to enter the foramen transversarium of the C6 vertebra. It can potentially be injured when releasing anterior scalene muscle while exposing proximal nerve roots.

Infraclavicular Region

Exposure of the supraclavicular and retroclavicular brachial plexus is adequate for exploration of the vast majority of brachial plexus injuries. If further exposure is needed, an extensile approach may include a deltopectoral incision. The cephalic vein is found in the interval between deltoid and pectoralis major. Depending upon the exposure needed, the deltoid and pectoralis major can be separated, the pectoralis major can be split along its fibers, the clavicular head of pectoralis major can be detached, or the entire pectoralis major can be taken down. The coracoid process is identified and pectoralis minor can be

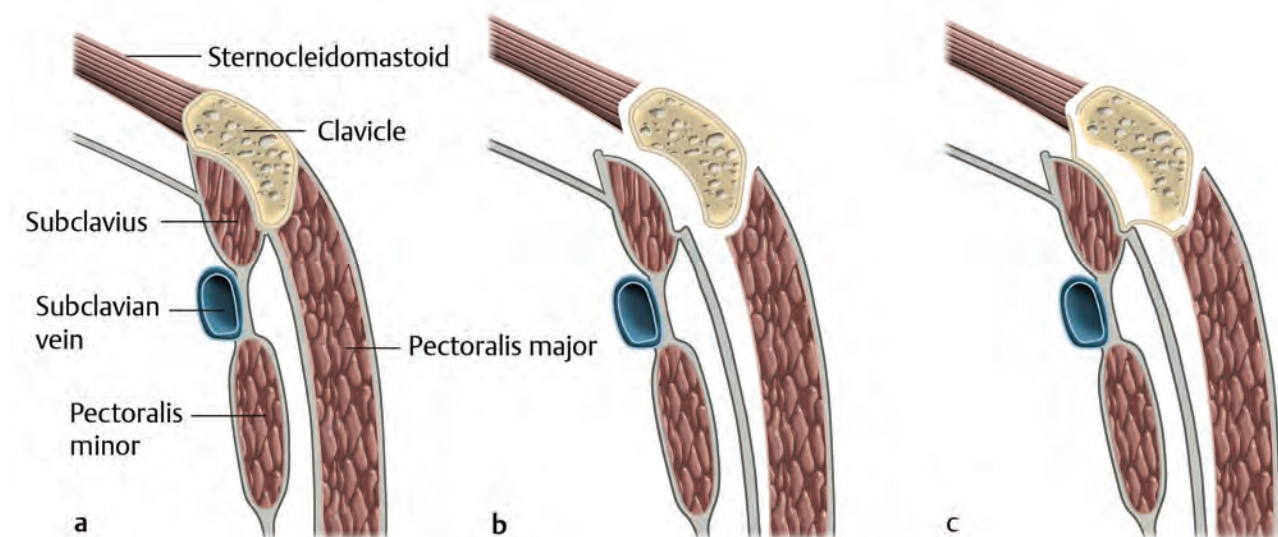


Fig. 38.10 Clavicle suspension. Periclavicular dissection for suspension (parasagittal section). **(a)** The sternocleidomastoid and pectoralis muscle originate from the cranial and caudal surfaces of the clavicle. The subclavius originates from a groove on the undersurface of the clavicle. Its fascia is continuous with the clavipectoral fascia and the fascia of the omohyoid. **(b)** Suprapariosteal dissection requires dissection of the subclavius from the groove in the clavicle, thereby providing a layer separating the suspending lace/tape from the subclavian vein. **(c)** Subperiosteal dissection can facilitate dissection of the subclavius from the groove in the clavicle. Incisions can be placed to preserve the anterior periosteum.

disinserted to reveal the underlying fat encompassing the neurovascular structures.

At this level, the most superficial element of the brachial plexus is the lateral cord, which gives off a lateral branch that becomes the musculocutaneous nerve (and runs through coracobrachialis) and a medial branch that contributes to the median nerve. The medial cord is found further medially and posterior to the axillary artery. The posterior cord and the axillary and radial nerve branches are best exposed with dissection lateral and posterior to the axillary artery. The axillary nerve branches first toward the quadrilateral space between the subscapularis and latissimus dorsi. The radial nerve crosses over the latissimus dorsi tendinous insertion before diving into the triangular space.

Clavicular Suspension

Retraction or elevation of the clavicle to open the costoclavicular space greatly facilitates the retroclavicular exposure. In order to reduce reliance on manual retraction, we suspend the clavicle with an umbilical tape that is passed around the clavicle in a subperiosteal plane to avoid the subclavian vein (► Fig. 38.10). The umbilical tape can be suspended on a retractor system such as a pediatric Bookwalter (► Fig. 38.11). Using this technique, we have been able to achieve satisfactory retroclavicular exposure without performing clavicular osteotomy.

38.5.3 Intraoperative Assessment of Brachial Plexus

Direct inspection of the brachial plexus provides insight into the location and extent of injury. A neuroma of the upper trunk

is frequently encountered (► Fig. 38.5), and proximal dissection to normal-appearing nerve roots is important to define the zone of injury and assure healthy proximal roots to graft from. When the middle trunk is injured (► Fig. 38.8), it is often encased in the scar of the upper trunk neuroma or ruptured. When lower roots are involved, avulsions are progressively more common at lower root levels (► Fig. 38.9).

Complete avulsions can be confirmed by one of the following:

- Finding a dorsal root ganglion outside of the bony foramen (► Fig. 38.9). The dorsal root ganglion can be confirmed on frozen section histology.
- Finding an empty bony foramen devoid of nerve tissue.
- Finding a normal-appearing nerve root (i.e., no signs of distal lesion) that is clinically nonfunctional on preoperative exam and does not respond to electrical stimulation. In this case, there is no neuroma formation as the injury is preganglionic.

Each nerve root is assessed by electrical stimulation. The voltage applied and the elicited motor response are recorded. Neurolysis has previously been performed as a treatment for nerve injury; however, studies have failed to demonstrate consistent clinical improvements using this strategy and the results of excision and grafting have been shown to be superior. Much of the decision to excise a portion of the brachial plexus is made based on preoperative clinical function, since a response to electrical stimulation alone does not necessarily indicate clinically useful motor function. The injured segment of nerve is excised to healthy-appearing nerve using loupe magnification.

The ends of the nerve are marked and submitted to pathology for frozen section histology. If a nerve end displays features of a neuroma (architectural disorganization, perineural

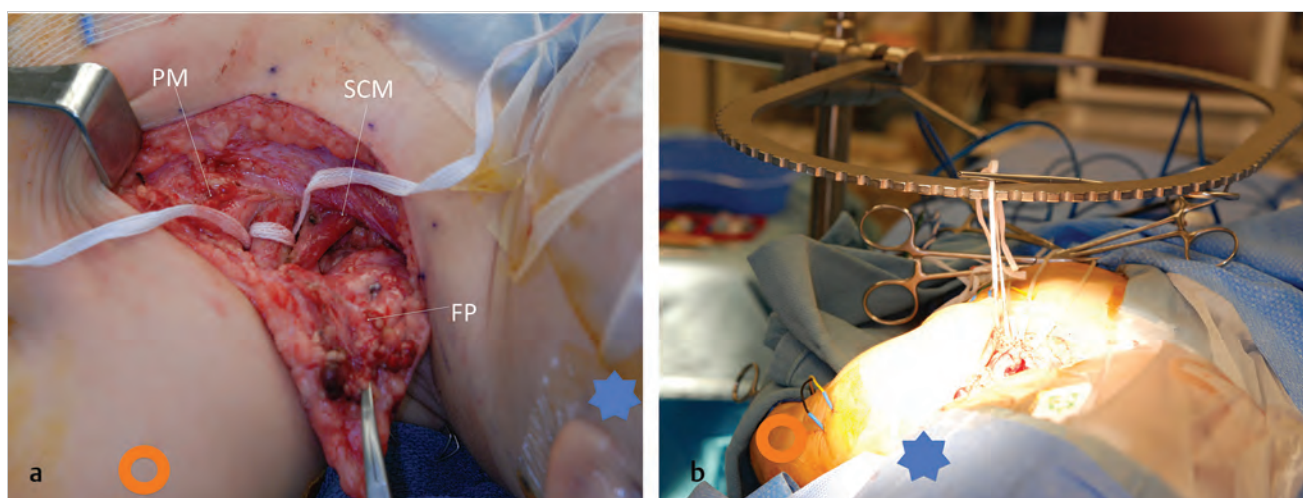


Fig. 38.11 Clavicle suspension. Supero-lateral view of left shoulder and neck. Blue star is on patient's face and orange circle is on patient's shoulder. (a) The superolateral-based supraclavicular fat pad (FP) has been turned over and both the sternocleidomastoid (SCM) and pectoralis major (PM) have been exposed and released. Circumferential dissection around the lateral portion of the clavicle allows umbilical tape to be passed around it. (b) Suspension apparatus. The left chest and shoulder arm are in view. A sterile stand is used for suspension of the clavicle.

microfascicles, or severe endoneurial/perineurial/epineurial fibrosis), the nerve can be cut further away from the zone of injury. Proximally, the bony foramen limits the extent in which a nerve can be trimmed. Distally, the nerve can generally be trimmed to healthy-appearing nerve. Some perineurial fibrosis of the distal target is accepted if there are no perineurial microfascicles because further trimming of the nerve results in longer grafts that may adversely affect reinnervation.

The combination of the histologic appearance of the proximal nerve roots and the response to electrical stimulation is used to determine the relative quality of proximal nerve roots for nerve grafting.

The use of intraoperative nerve conduction studies and of evoked potentials have been reported; however, there is disagreement with regard to the use of these studies and their correlation to clinical outcomes.

38.5.4 Reconstruction for Neonatal Brachial Plexus Palsy

Depending on the clinical scenario and intraoperative findings, specific reconstructive strategies will vary. There is general consensus that reconstructive priorities should be:

- Hand function.
- Elbow flexion.
- Shoulder stability and function.

Excision and grafting are the current standard for nerve reconstruction. Although nerve transfer as a primary strategy for reconstruction has gained popularity in the treatment of adult brachial plexus palsy, the role in treatment of NBPP has yet to be fully defined. Results of reconstruction using nerve transfers have been reported; however, these are limited to case reports and small case series. Nerve grafts take advantage of the abundant proximal regenerating axons, allow for anatomic reconstruction, and avoid sacrifice of motor donors. Nerve transfers

reduce the distance of regenerating axon to target, allow for greater motor-to-motor specificity, require fewer sites of coaptation, and can be technically easier to perform. There are no direct comparisons of nerve transfers to nerve grafting as a sole strategy for reconstruction of NBPP, so the relative efficacy remains unclear (Tse 2015). It is possible that multiple nerve transfers for Erb/type 1 palsies may have similar results to nerve grafting; however, when more than the upper plexus (C5–C6) is involved, nerve transfers alone will be inadequate for maximal reinnervation. Reinnervation of as many targets as possible provides the best outcomes and options for secondary reconstruction. Any surgeon engaging in nerve reconstruction for NBPP should be willing and able to perform nerve exploration and graft reconstruction as well as nerve transfers.

There may be a number of reasons that the relative advantage of nerve transfers in adults may not be the case in infants. Compared to adult brachial plexus palsy, infants with NBPP greatly differ in the mechanisms, patterns, severity, extent of injury, and scar tissue formation. The potential for neuromuscular recovery diminishes with age, and infants have a shorter limb over which axons need to regrow to reach their targets. In addition, due to rapid growth and development, infants possess much greater potential for central nervous system adaptation.

Although the role of nerve transfers as a sole strategy for primary reconstruction of Erb (Type I) brachial plexus palsy is unclear, there are several specific situations in which nerve transfers are preferred over nerve graft reconstruction. These situations include the following:

- Inadequate proximal nerve roots (i.e., avulsion).
- Delayed presentation.
- Isolated deficits.
- Failed primary reconstruction.

The following clinical scenarios illustrate some principles in the selection of strategies of nerve graft and nerve transfer for reconstruction of NBPP.

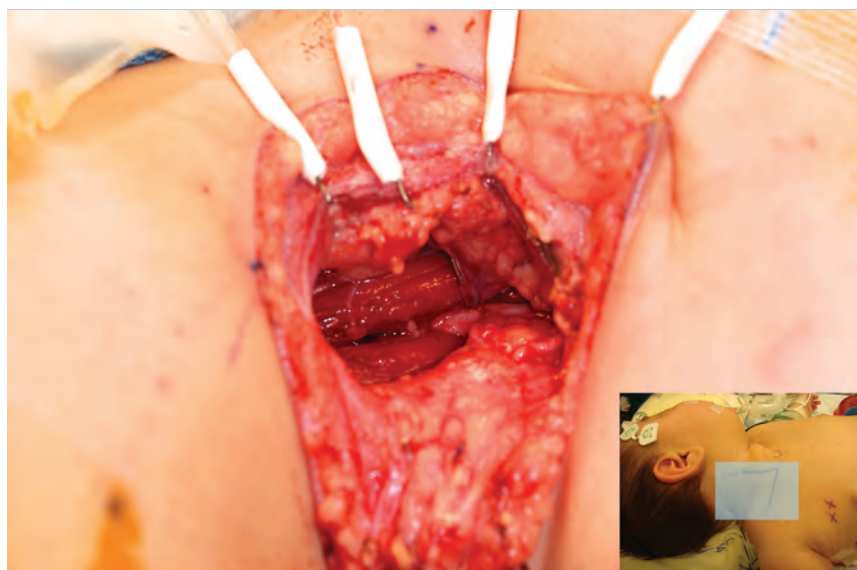


Fig. 38.12 Nerve graft reconstruction: The neuroma of the upper trunk has been excised in preparation for grafting.



Fig. 38.13 Nerve graft reconstruction. Bilateral sural nerve grafts are harvested via three short transverse incisions. Incisions are planned along creases at the ankle and popliteal fossa. The middle incision is planned at the junction of muscular and tendinous gastrocnemius.

Neonatal Brachial Plexus Palsy Example 1: Upper Plexus Palsy (Isolated Neuroma of Upper Trunk)

In the case of an upper trunk neuroma (► Fig. 38.5) and when both C5 and C6 roots are available, axons are delivered to target by cable grafts. The neuroma is excised in preparation (► Fig. 38.12). Bilateral sural nerve grafts are harvested under direct visualization via three small transverse incisions

(► Fig. 38.13). If visualization is difficult, an endoscope is used. The first priority of reconstruction is elbow flexion, followed by shoulder control. A surgical plan is devised based upon preoperative clinical function, findings on exploration, direct electrical stimulation of the proximal nerve roots, and histologic examination of cut ends to determine the relative quality of proximal stumps (► Fig. 38.14).

On the recipient side, the SSN innervates supraspinatus and infraspinatus (shoulder abduction and external rotation, respectively). The PD-UT is a major contributor to the axillary nerve that innervates the deltoid (shoulder flexion and abduction). The AD-UT is a major contributor to the musculocutaneous nerve that innervates the primary flexors of the elbow.

The cranial surface of the proximal C5 stump corresponds to the fascicular group of SSN and is generally reserved for this purpose. The cross-sectional surface of C5 can accommodate two additional sural nerve grafts, and these are used to innervate AD-UT and PD-UT. C6 tends to be a larger root and has a cross-sectional area that fits four sural nerve grafts. Two can be directed to AD-UT and two can be directed to the PD-UT. One advantage of this approach is if one of the two roots happens to have poor axon sprouting (i.e., unrecognized proximal injury), the crossover from roots may avoid total failure (► Fig. 38.14).

An alternate approach is to arrange the cable grafts into a more anatomic configuration with two grafts from C5 to the posterior division, a single graft from the cranial portion of C6 to the posterior division, and the remaining three grafts from C6 to the anterior division. A disadvantage of this approach is a lack of crossover between roots, meaning a higher chance of failure if there is an unrecognized proximal injury to one of the roots.

Nerve coaptation is performed under microscope securing grafts with fibrin glue proximally (► Fig. 38.15) and then distally (► Fig. 38.16), working from deep to superficial. The fibrin should be allowed to set before releasing retractors to avoid adherence and potentially tethering to surrounding tissues (► Fig. 38.17).

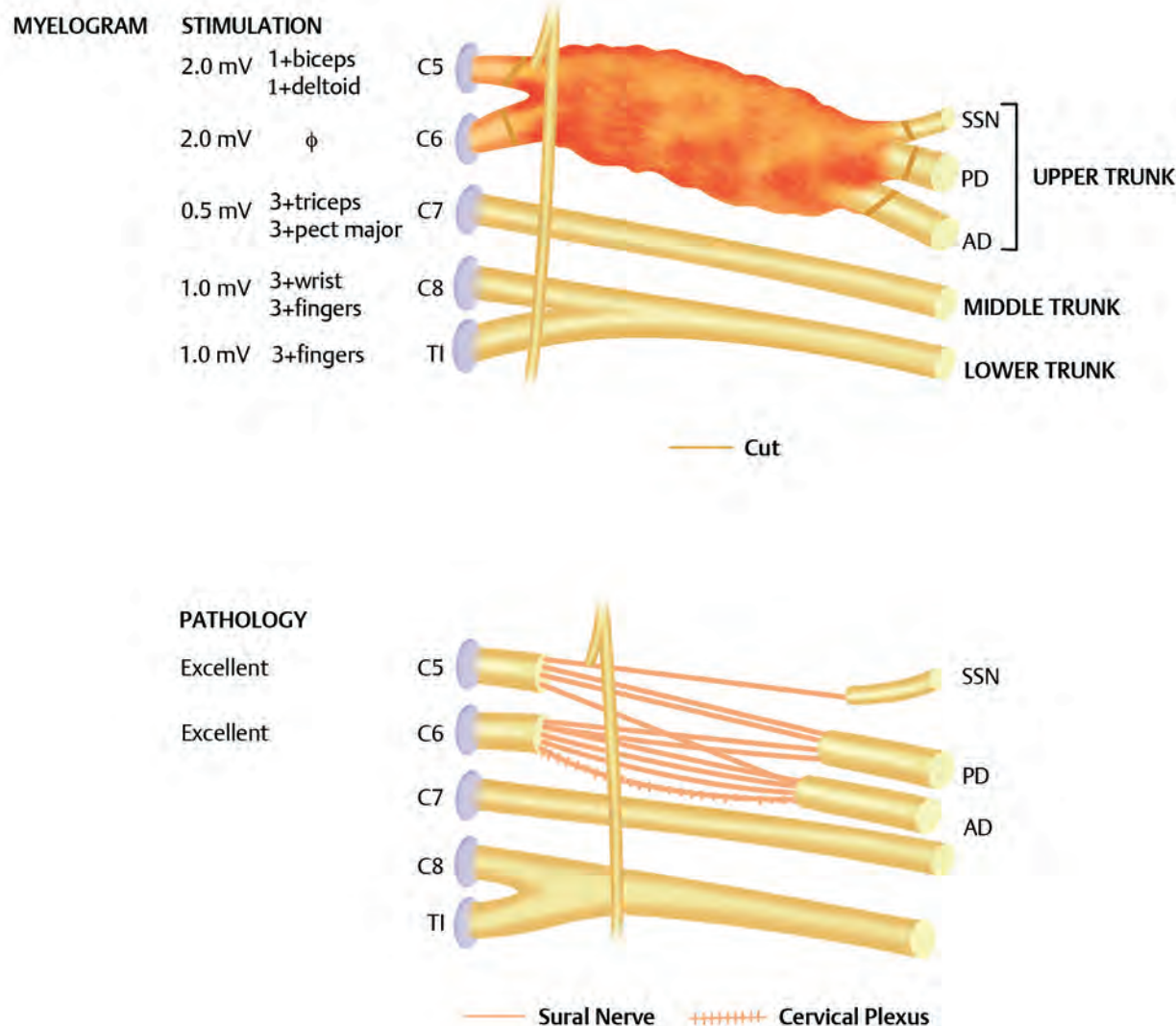


Fig. 38.14 Example case 1, surgical note (upper trunk neuroma): findings and plan for reconstruction. Neuroma is resected and cable grafts are used to innervate suprascapular nerve (SSN), posterior division of upper trunk (PD-UT), and anterior division of upper trunk (AD).

Neonatal Brachial Plexus Palsy Example 2: Upper Plexus Palsy (Lesions of Upper and Middle Trunk)

The strategy in this scenario is similar to example 1 with the addition of grafts from C7 to middle trunk (► Fig. 38.18). Sural nerve grafts are preferentially used to reinnervate the upper trunk and restore elbow flexion via AD-UT and shoulder abductors via PD-UT and SSN. Given the redundancy of function, the lower quality cervical plexus grafts are used for the middle trunk.

Neonatal Brachial Plexus Palsy Example 3: Pan Plexus Palsy with Two Avulsions (C8–T1)

In the case of pan-plexus palsy, reinnervation of the hand is the first priority (► Fig. 38.19). The intrinsic hand muscles and

extrinsic finger flexors are targeted via the T1 and C8 roots of the lower trunk. The best-quality proximal roots and nerve grafts are dedicated to these targets. In the case of a nerve root avulsion in which the dorsal root ganglion is found in the supraclavicular space, neurolysis can be performed to separate the dorsal root from the ventral root. In doing so, the sensory and motor nerve components, respectively, can be isolated. The lower trunk can then be mobilized so that a direct coaptation between C7 and the ventral root (pure motor component) of the T1 target can be achieved. When the ventral and dorsal roots cannot be separated, as with C8 in this case, short grafts are used.

Elbow flexion is the next priority and is targeted via grafts to AD-UT. The shoulder is the third priority and is targeted via grafts to PD-UT with an additional nerve transfer to SSN. Given that two of three roots in this case are not available, the number

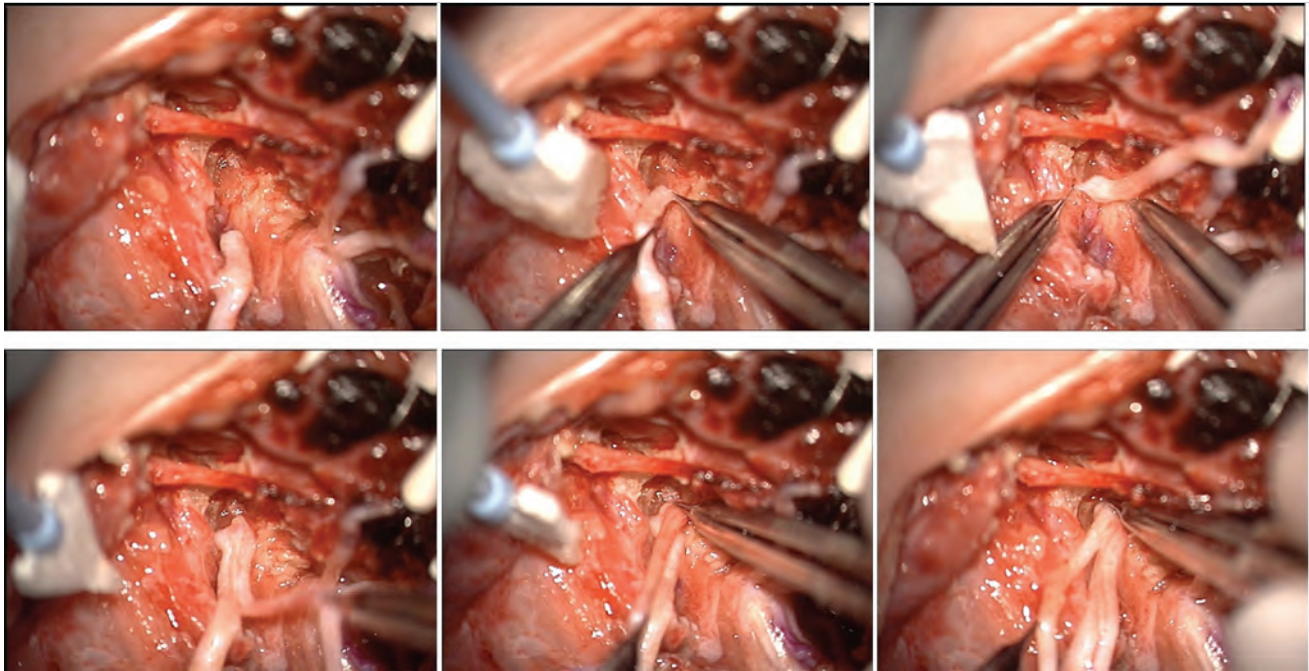


Fig. 38.15 Nerve graft reconstruction. View through microscope of cable graft coaptations to proximal C5 nerve root at the intervertebral foramen. The C6 graft inset can be seen at bottom right of each frame.

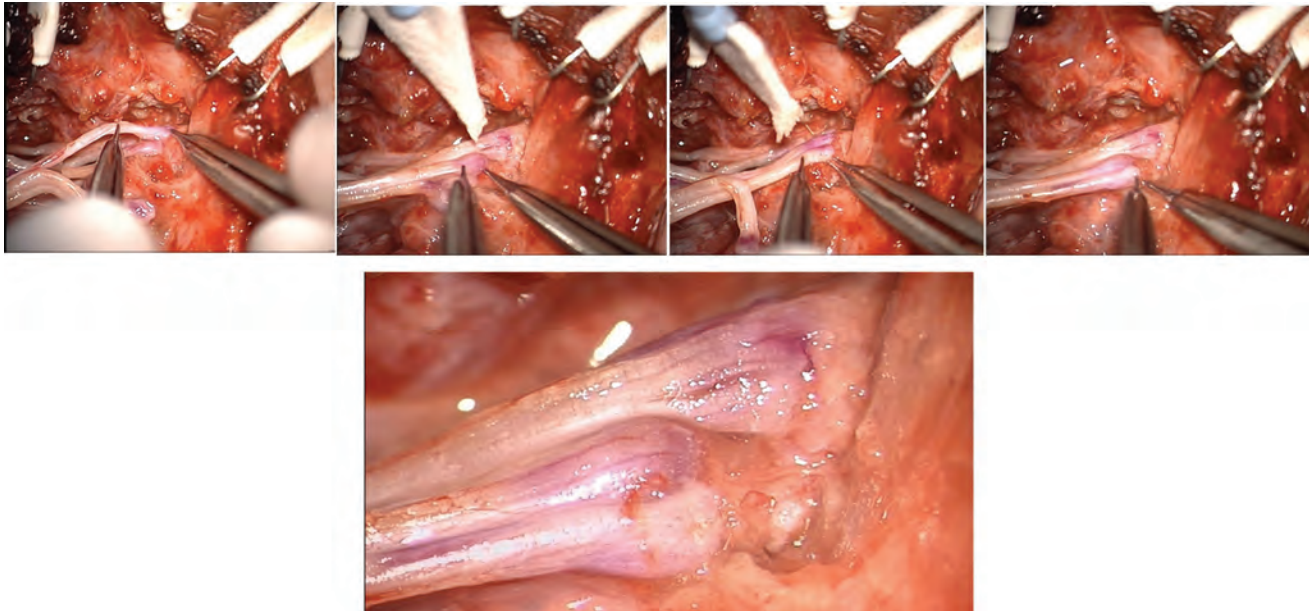


Fig. 38.16 Nerve graft reconstruction. Coaptation of nerve grafts to distal target nerve stumps.

of axons for potential reinnervation is reduced. The spinal accessory nerve (SAN) is a close match in size and axons to SSN and is thus used as a donor for transfer.

The remaining functions of the brachial plexus are the last priority, and the lower quality cervical plexus graft is typically used to target the middle trunk.

Neonatal Brachial Plexus Palsy Example 4: Pan Plexus Palsy with Three Avulsions (C7–T1)

Meaningful function can still be achieved in spite of three avulsions (► Fig. 38.20). Hand function is the first priority, and three grafts are dedicated to the lower trunk. Two grafts are used to

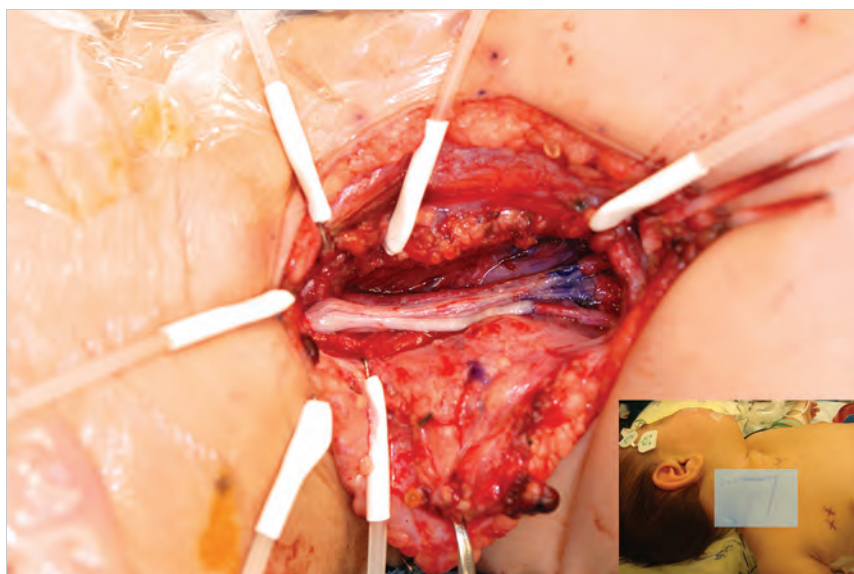


Fig. 38.17 Nerve graft reconstruction. Multiple cable grafts secured using fibrin glue according to surgical plan (► Fig. 38.14).

target AD-UT and PD-UT for elbow flexion and shoulder control. Additional axons are recruited with a SAN-SSN transfer. The remaining cervical plexus graft is used for the lowest priority target, the middle trunk.

Neonatal Brachial Plexus Palsy Example 5: Pan Plexus Palsy with Four Avulsions (C6–T1)

When four of five roots are avulsed, an additional source of axons is needed to restore meaningful motor function (► Fig. 38.21). In this case, both C8 and T1 roots are isolated and two grafts are dedicated to each for reinnervation of the hand. SAN-SSN transfer and grafts to PD-UT address the shoulder. Additional axons for innervation can be obtained from the third, fourth, and fifth intercostals (see section “Intercostal to Musculocutaneous Nerve Transfer”), which are transferred directly to the musculocutaneous nerve with a coaptation in the axilla.

Neonatal Brachial Plexus Palsy Example 6: Upper Palsy with Delayed Presentation or C5 and C6 Avulsions

In the case of breech presentation, there may be an upper palsy with C5 and C6 avulsions and no proximal nerve roots available for grafting. In the case of delayed presentation, moving the site of nerve repair closer to the distal target allows for earlier reinnervation.

Multiple nerve transfers are indicated in these cases (see section “Technical Notes for Nerve Transfers”). In addition to SAN-SSN, one of the three motor branches of radial nerve to triceps can be transferred to the axillary nerve (Tri-Ax). Reinnervation of elbow flexion can be achieved via a bifascicular transfer (M/U-Bi/Br), in which a redundant fascicle of ulnar (FCU) nerve is transferred to the brachialis branch of musculocutaneous nerve, and a redundant fascicle of median nerve (flexor digitorum superficialis [FDS] or flexor carpi radialis [FCR]) is transferred to the biceps branch. We prefer this double fascicular transfer in these situations, in which a greater number of motor axons are

transferred for elbow flexion restoration than with the traditional single ulnar to musculocutaneous transfer originally described by Oberlin.

Neonatal Brachial Plexus Palsy Example 7: Upper Plexus Palsy with Isolated Deficits

Isolated deficits following brachial plexus palsy are unusual. In the case of poor elbow flexion with excellent shoulder abduction, the decision can be made to proceed with nerve graft reconstruction of the upper trunk with the goals of improving elbow flexion but with the accepted risk of downgrading shoulder abduction.

The alternative is to perform specific nerve transfer(s) to address the isolated deficit(s). The example illustrated in ► Fig. 38.22 was a 9-month-old who failed the Cookie test but who had excellent shoulder abduction. Elbow flexion was poor (less than 90 degrees against gravity with no progression over 6 months) and there was no shoulder external rotation. Preoperative myelogram was suggestive of an avulsion of C6. On exploration, C5 and the upper trunk had grossly recognizable architecture following neurolysis, but the C6 root was severely scarred up to the level of the upper trunk. An intact C5 contribution to upper trunk with an avulsion of C6 explained the clinical presentation. Given the excellent shoulder abduction and a lesion that may not be improved by excision and grafting, isolated nerve transfers were performed including SAN-SSN and M/U-Bi/Br.

Neonatal Brachial Plexus Palsy Example 8: Pan Plexus Palsy with Failed Reconstruction of Elbow Flexion

These situations are rare, and there is no consensus on the indications for secondary reconstruction. Clarke reported that recovery following NBPP plateaus at 2 to 3 years. Given this long time span and the competing threat of permanent motor endplate atrophy, it is difficult to determine when and if to intervene. In the case of nerve graft reconstruction for pan-

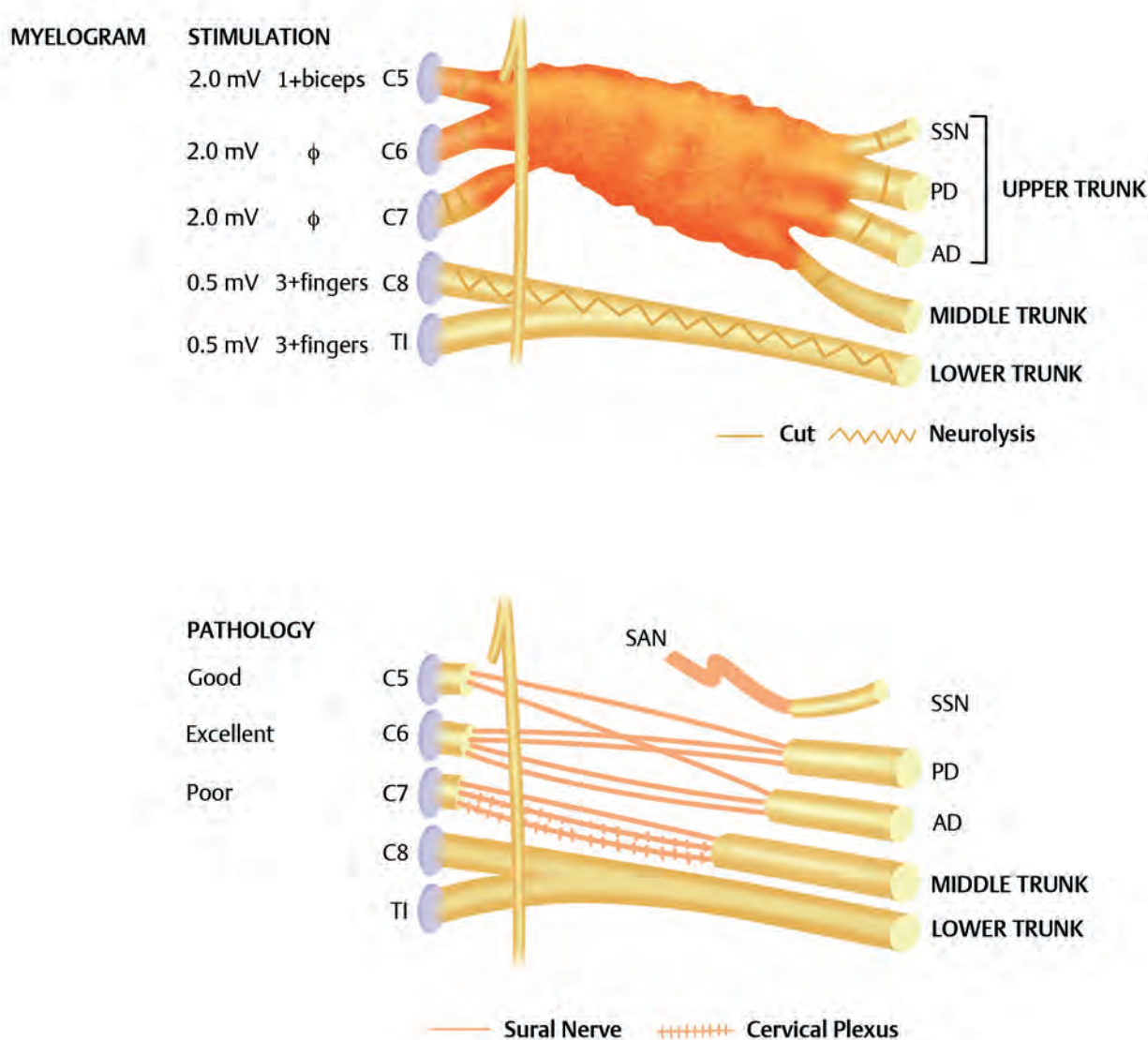


Fig. 38.18 Example case 2, surgical note (upper trunk neuroma, middle trunk rupture): findings and plan for reconstruction that include nerve grafts to posterior division of upper trunk (PD-UT) and anterior division of upper trunk (AD). The suprascapular nerve (SSN) was innervated by the spinal accessory nerve (SAN).

plexus palsy (as in NBPP example 4), if there is inadequate elbow flexion but adequate hand and shoulder function 1 year after surgery, it may be reasonable to offer secondary reconstruction. Intercostal (third, fourth, and fifth) to musculocutaneous nerve transfer is one option (see "Intercostal to Musculocutaneous Nerve Transfer"). In the author's experience of two cases, recovery of meaningful elbow flexion occurred 3 to 6 months after secondary reconstruction. It is possible that this recovery could still have occurred with primary reconstruction alone, and parents must be informed that recovery without further surgery is possible. If secondary reconstruction is deferred and no meaningful elbow flexion is recovered, innervated free muscle transfer or other secondary musculoskeletal procedures remain options.

38.5.5 Reconstruction for Traumatic Brachial Plexus Palsy

Principles of TBPP reconstruction are similar to NBPP, but avulsions are more common and the distance from regenerating axon to target is greater. Similar to adults, older children have less regenerative capacity than infants. Nerve grafts may be used for reconstruction of proximal targets (i.e., shoulder) to take advantage of the abundant regenerating axons, but nerve transfers may be required for more distal targets (i.e., elbow) to bring regenerating axons closer to target. As has been reported in the treatment of adult brachial plexus palsy, the combination of nerve grafts and transfers likely maximizes potential outcomes.

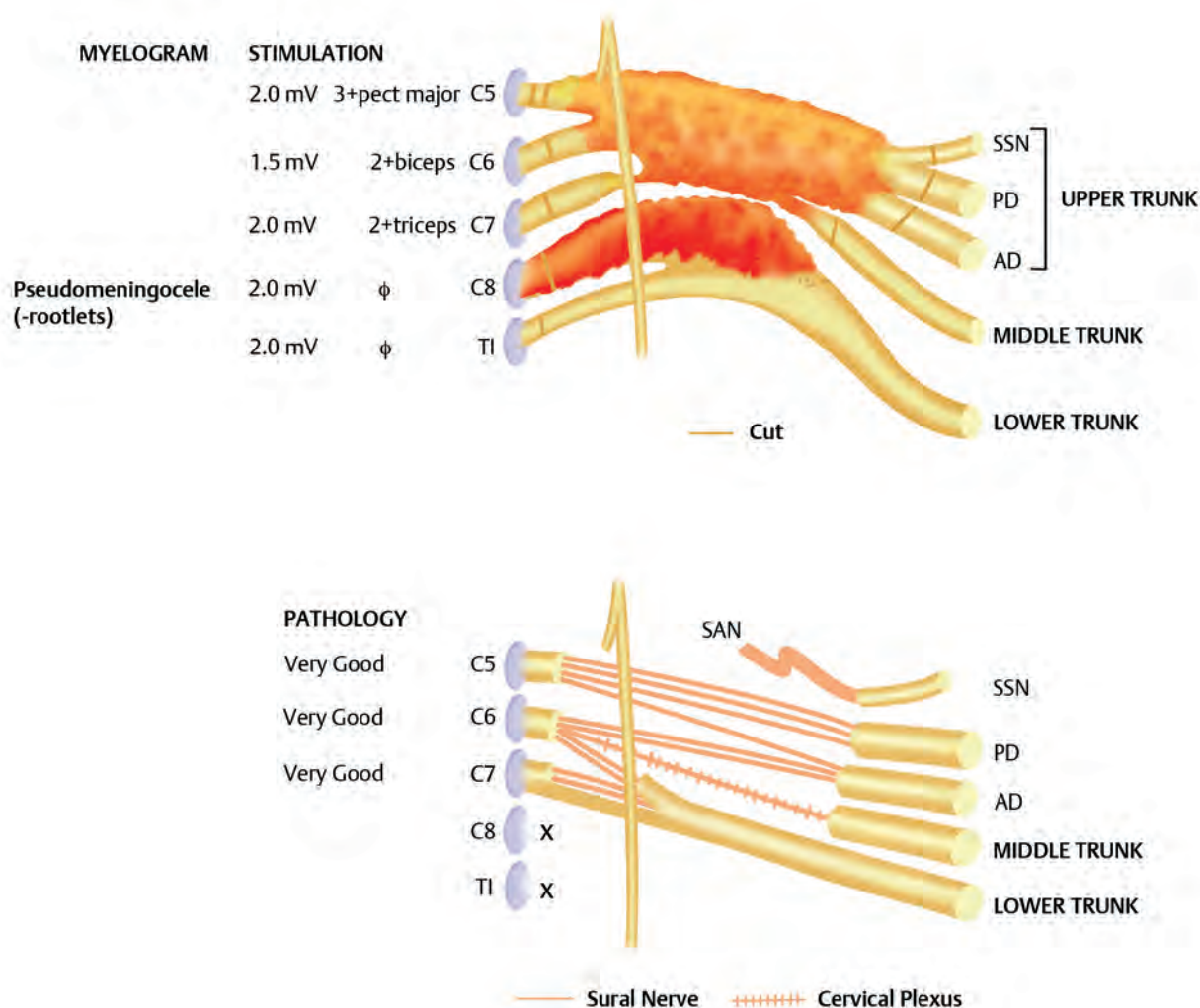


Fig. 38.19 Example case 3, surgical note (pan plexus palsy with two avulsions): findings and plan for reconstruction. On clinical examination, the hand was flail with no apparent function. On exploration, C8 was found to have progressively greater scarification that extended proximal to the vertebral foramen and T1 had a normal appearance with no response to electrical stimulation. The findings for both C8 and T1 were consistent with avulsion of rootlets from spinal cord. The C8 contribution to lower trunk was innervated via nerve grafts. The T1 contribution to lower trunk was repaired directly to the C7 stump.

38.5.6 Technical Notes for Nerve Grafting

Fibrin sealant is used as a “nerve glue” to ensure accurate end-to-end coaptation and optimize nerve recovery. Sutures have been found to produce granulomas that can impair nerve recovery. Sutures repair is also not possible if a proximal nerve root is trimmed to or within the bony intervertebral foramen (► Fig. 38.15). Grafts should be tailored in the position that requires the greatest nerve length (i.e., shoulder adducted and head turned to contralateral side) and with some redundancy to avoid any tension and to allow nerve gliding with movement. An organizational strategy helps with the management of multiple cables.

Each sural nerve is individually repaired under the microscope starting on the proximal side, working from the deepest

root/region to the most superficial (► Fig. 38.15 and ► Fig. 38.16). Although the internal neural topography of the brachial plexus is not well described, cables are organized on the proximal roots that mimic their course to distal targets. Dilator forceps can be used to manipulate epineurium and ensure accurate coaptation. The two components of fibrin sealant (i.e., fibrin and activator) can be delivered via separate syringes to avoid the delivering catheter from getting clogged when multiple applications are performed. Fibrin glue is only applied to the sites of coaptation so that the rest of the nerve graft can glide across tissues in the case of any movement.

Heat from the operating microscope can easily result in desiccation of nerve grafts, and significant soft-tissue burns have been reported in infants. Room lights are dimmed so the microscope light source can be lowered in intensity. The wound and nerve grafts should be irrigated frequently.

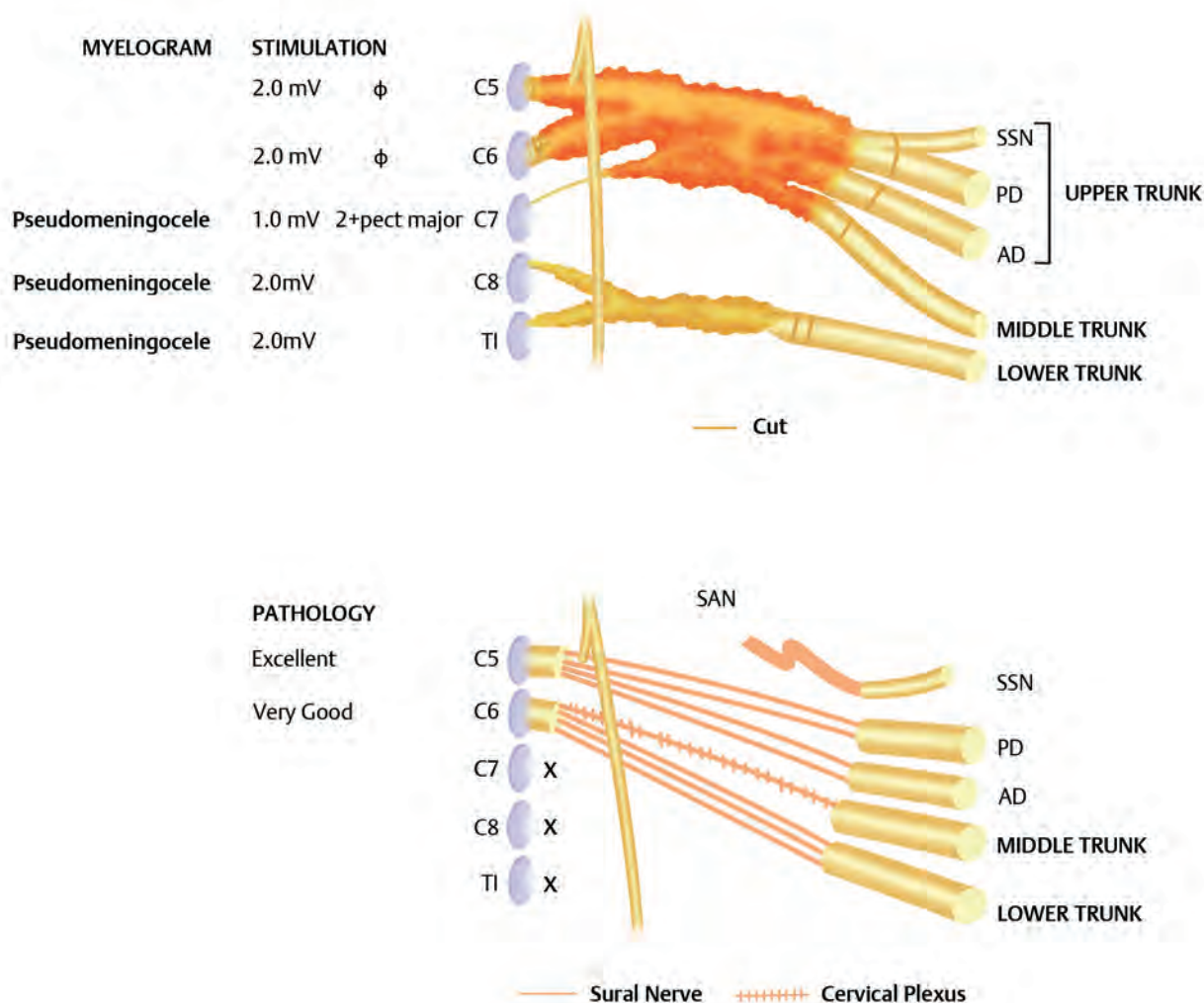


Fig. 38.20 Example case 4, surgical note (pan plexus palsy with three avulsions): findings and plan for reconstruction.

38.5.7 Technical Notes for Nerve Transfers

Donor nerve selection should be determined preoperatively by clinical exam and with EMG when needed (i.e., the donor may be partially innervated or have incomplete reinnervation). Intraoperative testing with an electrical stimulator is helpful in confirming adequate motor response of the donor nerve. The use of smaller voltages (0.1–0.5 mV) avoids overstimulation and electrical spread. Intermittent stimulation prevents tetanic contraction, which can result in fatigue. Donor nerves should have a strong response to stimulation, but caution must be employed to avoid sacrificing critical functions. Stimulation of nerves, branches, or fascicles being preserved is important to confirm that the critical function is left intact.

Nerve repair is best performed with redundancy in length, allowing for motion without consequent tension on the site of coaptation. To ensure length, the donor should be transected distally, while the recipient should be transected proximally. Neurolysis provides additional length.

Spinal Accessory to Suprascapular Nerve Transfer

When performed from the anterior approach, the SAN can be found between the supraclavicular fat pad and the trapezius muscle in the lower one-third of the neck. The nerve can be followed to the lower scapula, transected distally, and then passed through an opening in the supraclavicular fat pad to the SSN. This may be performed during primary nerve exploration and grafting. It is critical to leave one to two branches of SAN to upper trapezius intact to avoid significant scapulothoracic morbidity and scapula instability.

The posterior approach offers the advantage of SSN decompression, but requires prone positioning. The trapezius is split at the level of the scapular spine and the SAN is identified approximately 40% of the distance from midline to acromion (► Fig. 38.23a, b, c). The SAN should be followed as far distal as possible to ensure length. The SSN is found along the anterior border of the supraspinatus, approximately halfway between the superior angle of scapula and acromion (► Fig. 38.23a). Palpation of the coracoid anteriorly helps orient the surgeon to the

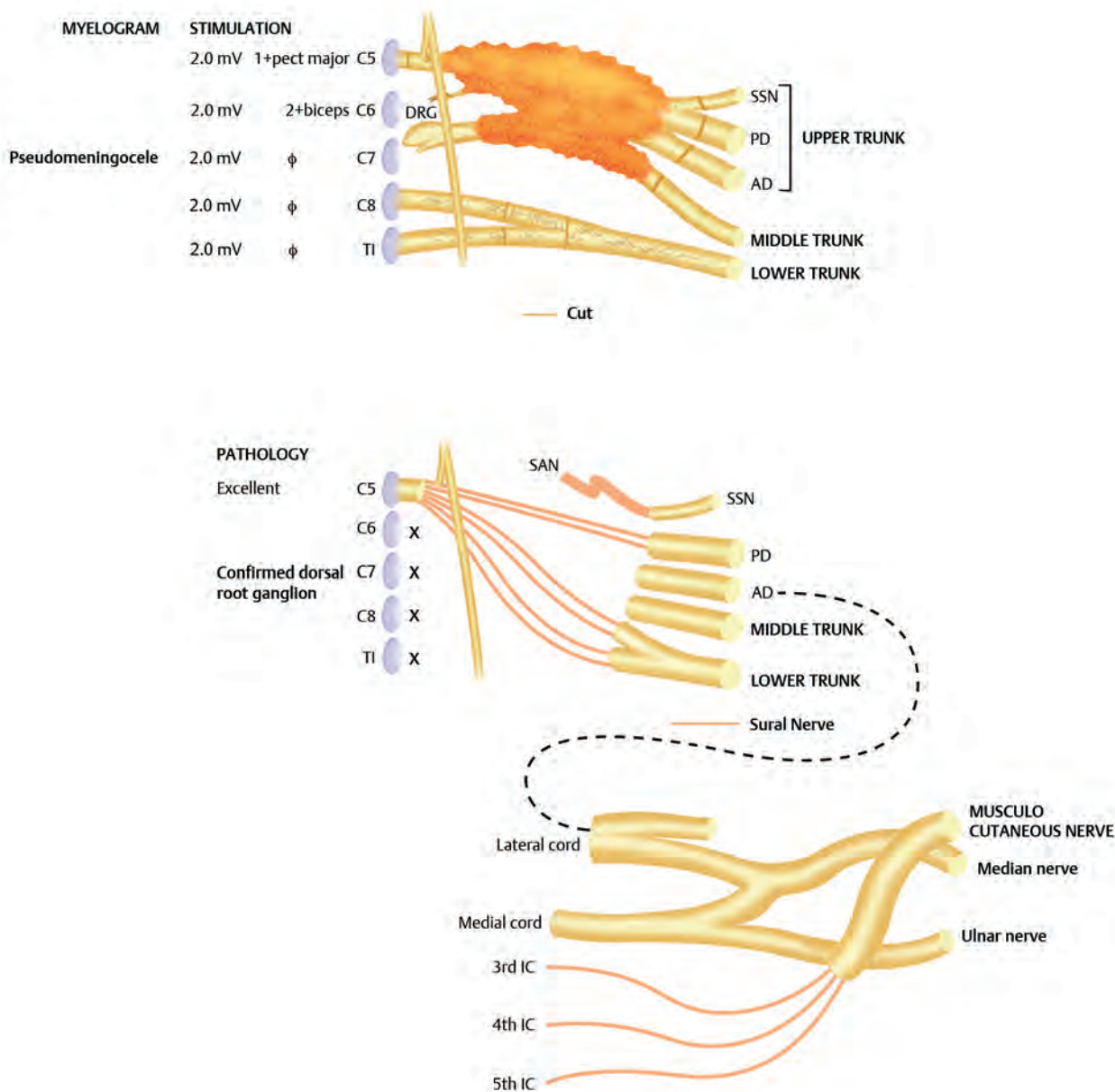


Fig. 38.21 Example case 5, surgical note (pan plexus palsy with four avulsions): findings and plan for reconstruction. Given the limited number of proximal roots to graft from, three intercostal nerves were used as a distal transfer to innervate musculocutaneous nerve, thereby targeting the elbow flexors.

vicinity of the scapular notch. Retraction of the scapula posteroinferiorly helps with the deep exposure. The origin of the omohyoid muscle can often be visualized and the adjacent notch palpated. The suprascapular artery can be seen coursing over the suprascapular ligament. Once this ligament is divided (► Fig. 38.23e), the nerve can be visualized and followed as proximal as possible to allow laxity for direct coaptation (► Fig. 38.23f).

Sacrifice of lower trapezius function bears little morbidity; however, the lower trapezius is sometimes used as a donor muscle for tendon transfer in the case of persistent deficits in

external rotation. When additional axons are needed for reconstruction, and if latissimus dorsi function is intact or to be reinnervated, sacrifice of the lower trapezius to potentially achieve external rotation is a reasonable strategy.

Triceps to Axillary Nerve Transfer

Preoperative EMG to examine the pattern and strength of voluntary innervation for each head ensures that an adequate donor is available and is helpful in the selection of an appropriate branch for transfer.

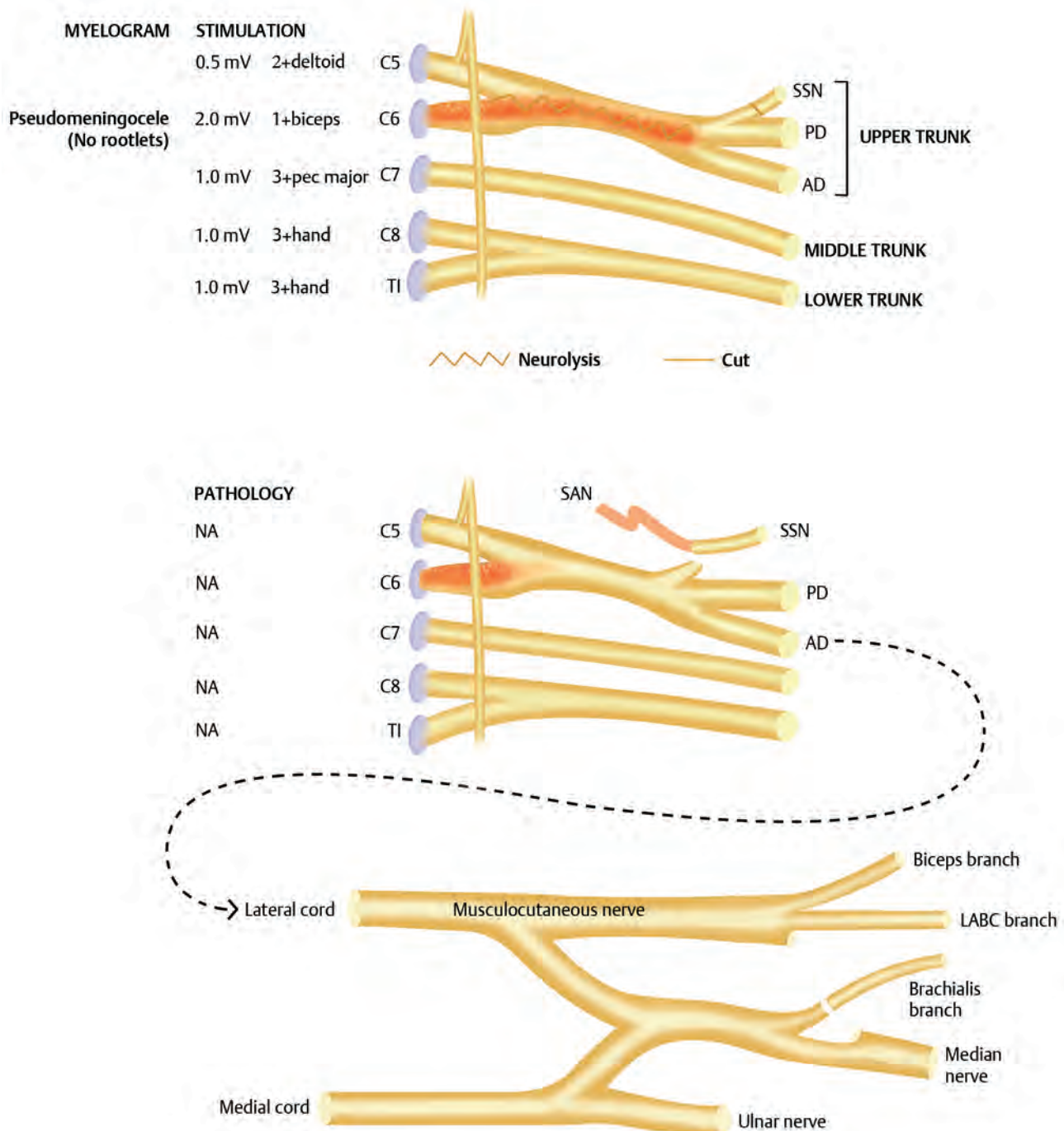


Fig. 38.22 Example case 7, surgical note (upper plexus palsy with isolated deficits): findings and plan for reconstruction. Given satisfactory shoulder abduction, specific deficits of shoulder external rotation and elbow flexion, and an apparent avulsion of the C6 root, the brachial plexus was left intact and distal nerve transfers were used for reconstruction. Spinal accessory nerve (SAN) was transferred to suprascapular nerve (SSN) and the flexor digitorum superficialis (FDS) fascicle of median nerve was transferred to brachialis branch of the musculocutaneous nerve.

The posterior approach provides ideal visualization to both donor and recipient branches. An incision is designed along the posterior midline of the arm extending to the posterior border of deltoid (► Fig. 38.24a). The interval between triceps and deltoid leads to the quadrangular space (► Fig. 38.24b, c). The sensory branch of the axillary nerve can then be traced to the posterior, anterior, and teres major branches (► Fig. 38.24d).

Depending upon the circumstances, the main trunk of the axillary nerve, or one of the posterior or anterior branches, can be selected as the target. Proximal dissection provides length for coaptation. The triangular space can be found on the inferior border of teres major, containing the branches to the three heads of triceps (► Fig. 38.24d). The branch to medial head is found overlying the radial nerve and is often larger with longer

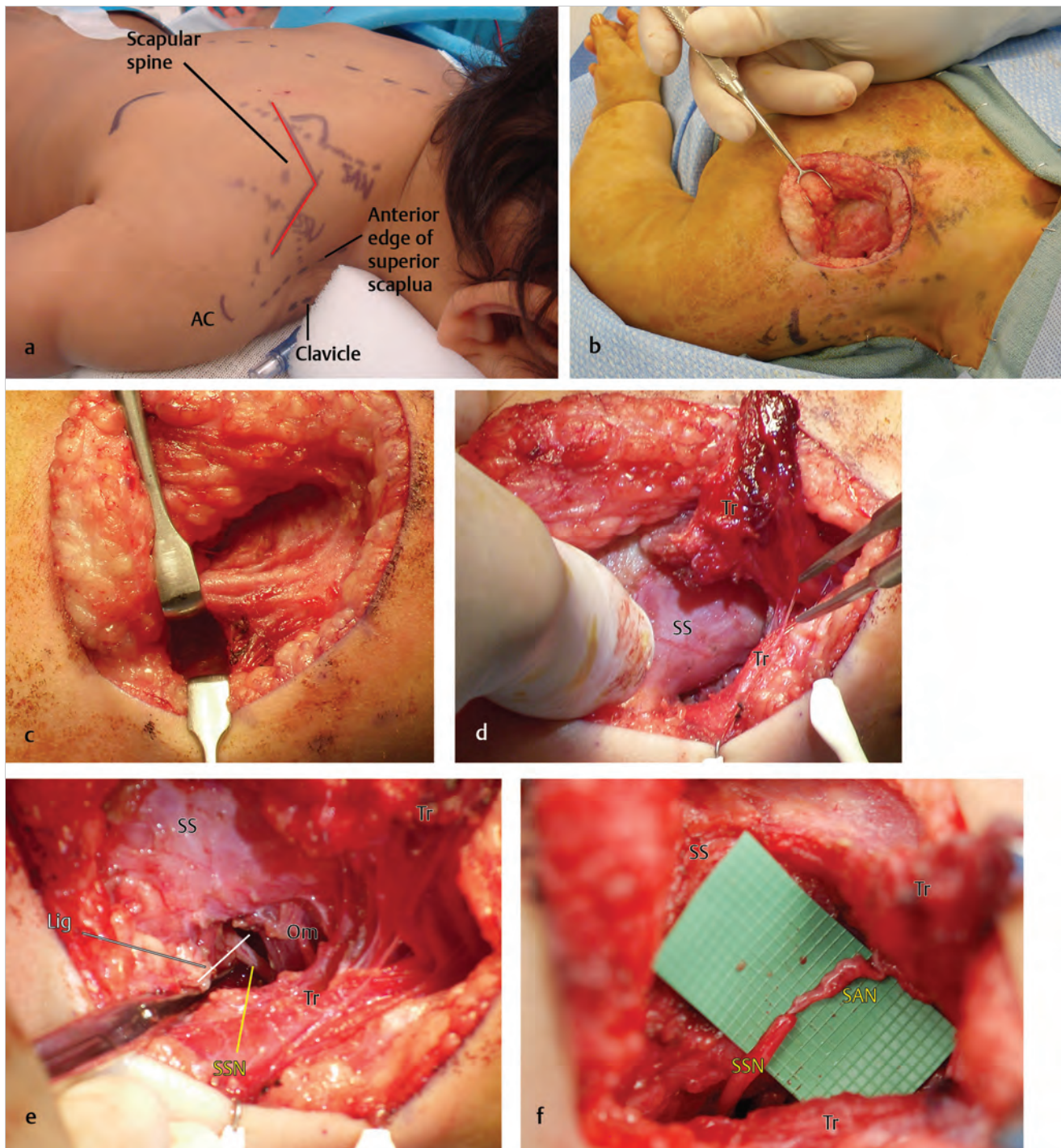


Fig. 38.23 Spinal accessory nerve (SAN) to suprascapular nerve (SSN) transfer. **(a)** Surface landmarks. The course of SAN is 40% of the distance between midline of the back and acromion (AC). The course of SSN is midway between superior angle of scapula and AC. The scapular spine and anterior edge of scapula define the borders of the supraspinatus muscle. **(b)** Exposure of trapezius. **(c)** Trapezius is split transversely between muscular fibers. **(d)** The trapezius (Tr) is also released along its bony insertion into the scapular spine. Supraspinatus muscle is exposed (SS). **(e)** Caudal traction on the scapular spine rotates the anterior edge of superior scapula into view. The suprascapular notch is difficult to visualize, but the origin of omohyoid (Om) helps define the medial edge of the notch. The SSN is visualized following release of the suprascapular ligament (Lig) and can be traced proximally (i.e., away from SS) up to the upper trunk to attain greater length for transfer. **(f)** The proximal stump of SSN is flipped posteriorly. The SAN is traced to the lower scapula before transection and flipping the stump anteriorly.

length that allows for redundancy. However, the ultimate donor branch selection is based on preoperative EMG and intraoperative examination using electrical stimulation (► Fig. 38.24e).

Fascicular Transfer for Elbow Flexion: Oberlin Type (M/U-Bi/Br)

Depending on the circumstances, either one or two fascicles can be transferred to either biceps or brachialis or both, respectively.

A medial arm incision is placed over the medial intermuscular septum. The deep muscular fascia anterior to the septum is incised so that the interval between biceps and brachialis can be opened (► Fig. 38.25a). The musculocutaneous nerve can be seen passing through the coracobrachialis before dividing into its branches (► Fig. 38.25b). The biceps branch is most proximal and travels lateral. The brachialis branch is next and travels medial (often obscured by a crossing vessel from brachial artery to biceps). The lateral antebrachial cutaneous (LABC) nerve continues and travels into the antecubital fossa. The three branches are lysed as far proximal as possible.

The medial antebrachial cutaneous (MABC) nerve is superficial and can be mistaken for a major motor nerve. The median nerve is found almost immediately medial to the musculocutaneous nerve, and the ulnar nerve is found further medially and deep to the brachial artery. Small segments of both the ulnar and the median nerve epineurium are opened to for internal neurolysis to expose appropriate donors (► Fig. 38.25b). Mackinnon has described the relevant topographical fascicular anatomy that allows appropriate fascicles to be isolated without

dissection of the rest of the nerve (► Fig. 38.25c). For the median nerve, FDS and FCR fascicles are found on the medial superficial surface of the nerve (the pronator fascicle is on the superficial central region, sensory fascicles are found on the lateral side, and the anterior interosseous nerve (AIN) fascicles are on the deep side of the nerve). For the ulnar nerve, the FCU branch is found on the lateral superficial aspect of the nerve. It is critical to avoid sacrifice of any flexor digitorum profundus (FDP) or intrinsic function. Appropriate planning ensures redundant nerve length for transfer (► Fig. 38.25d).

Electrical stimulation should be used to confirm that the fascicle being sacrificed has redundancy of function in the fascicles being left behind. Loss of hand function in a compromised limb is a risk and can be devastating. The effects of median and/or ulnar fascicle sacrifice on hand growth seem to be insignificant in the short term; however, long-term studies are not yet available.

Intercostal to Musculocutaneous Nerve Transfer

The intercostal nerves contain both motor and sensory components. When used as donors for elbow flexion, three levels are generally needed to provide adequate axons. The line of the anticipated inframammary fold is drawn and an incision is designed along a short segment of this (► Fig. 38.26a). The lateral border of the pectoralis major is identified and elevation exposes the chest wall (► Fig. 38.26b). Fibers of pectoralis minor and serratus anterior must be split to expose the anterior surface of the third, fourth, and fifth ribs. The anterior periosteum is incised and dissected from the lower border and

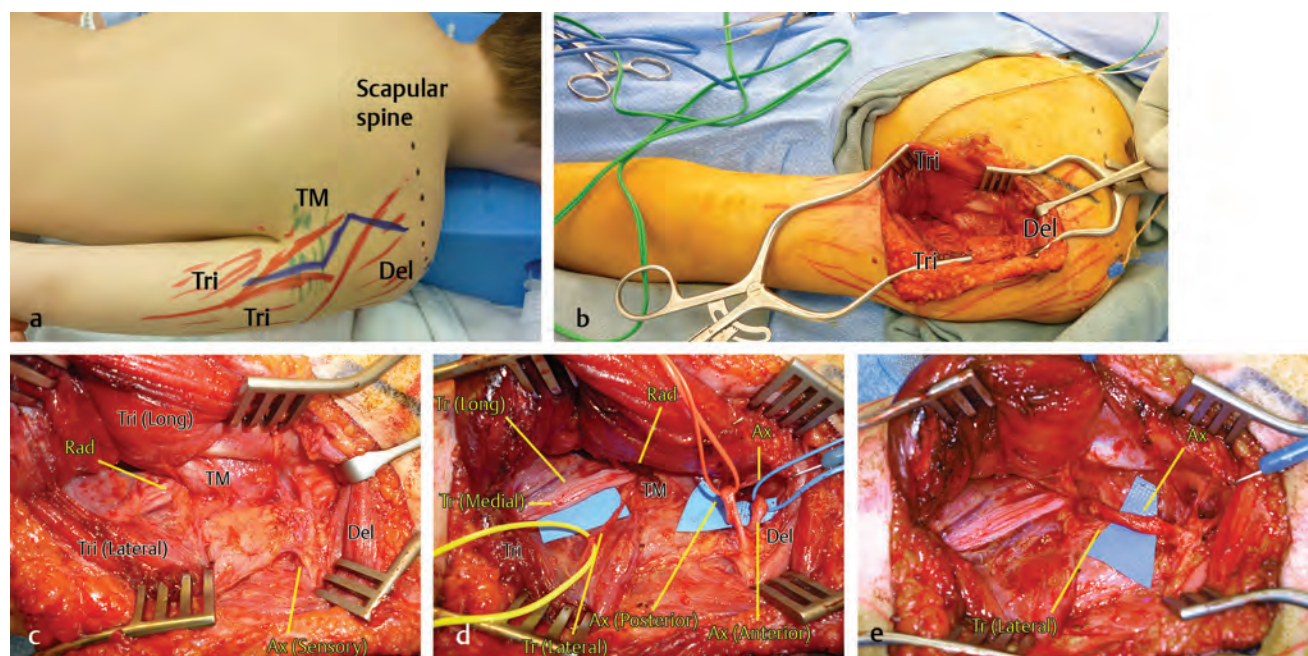


Fig. 38.24 Triceps branch of radial nerve (Tr) to axillary nerve (Ax) transfer. (a) Surface landmarks. The interval between deltoid (Del) and the heads of triceps (Tri) can be palpated. The anticipated location of teres major (TM) is marked. (b) The interval is opened. (c) The radial nerve (Rad) exits the triangular space inferior to teres major, whereas the axillary nerve (Ax) exits the quadrangular space superior to teres major. The sensory branch of axillary nerve can also be followed toward to the quadrangular space. (d) Branches of radial and axillary nerve are identified. The axillary branches can be followed proximally to the nerve trunk. (e) The donor triceps branch is followed distally to allow the nerve to swing superiorly. The axillary nerve is dissected proximally to swing the stump inferiorly.

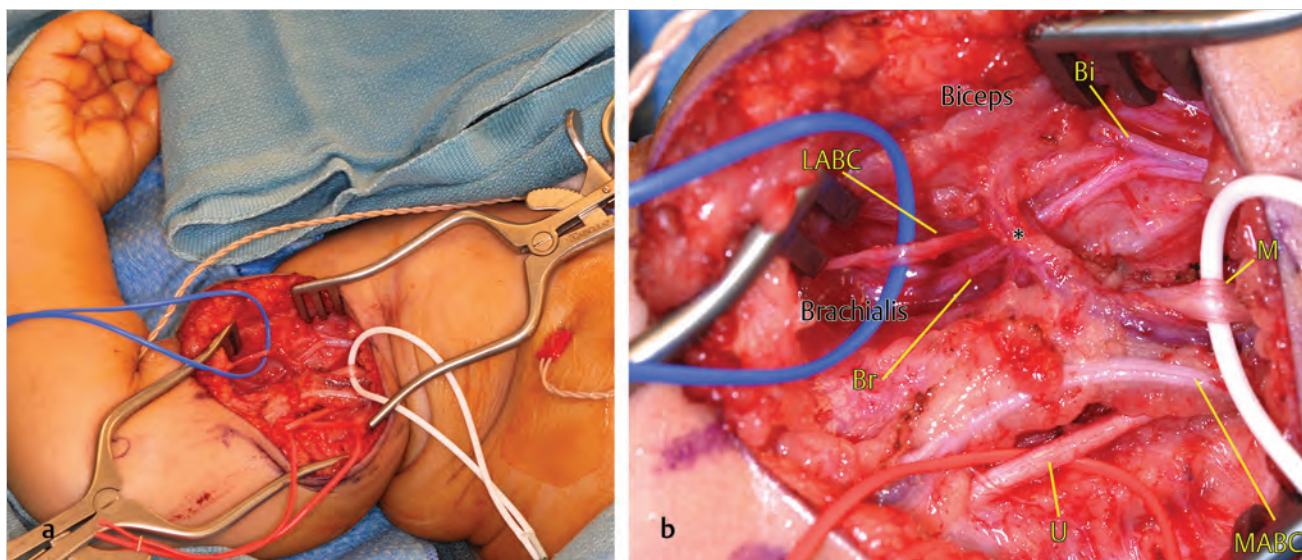


Fig. 38.25 Fascicular transfer for elbow flexion (M/U-Bi/Br). (a) Incision along medial arm and through muscular fascia to expose the space between biceps and brachialis. (b) The branching pattern of musculocutaneous nerve is consistent. The biceps branch (Bi) is lateral, the brachialis branch (Br) is medial, and the lateral antebrachial cutaneous (LABC) nerve continues its central course. A vascular branch of brachial artery to biceps crosses the divergence of brachialis branch from the LABC. Medial antebrachial cutaneous nerve (MABC) is found superficial to the brachial neurovascular bundle. Short segments of median (M) and ulnar (U) nerve are exposed for transfer. (Continued)

posterior surface of the rib, thereby allowing downward retraction of the intercostal muscle and neurovascular structures away from bone (► Fig. 38.26c). Incision through periosteum and gentle dissection through intercostal fibers reveal the intercostal nerve. There is often only a thin layer of tissue overlying pleura. The nerve can be followed medially to the costochondral junction before motor function diminishes, and then mobilized posteriorly, which involves sacrificing the lateral cutaneous branch for sufficient length. A tunnel through the serratus anterior muscle provides a direct course toward the axilla.

The musculocutaneous nerve is identified in the upper medial arm and followed proximally under the pectoralis major to its origin from the lateral cord in order to provide length (► Fig. 38.26d). The coracobrachialis often must be sacrificed to allow direct coaptation to donor nerves in the axilla.

Sacrifice of intercostal nerves in the setting of ipsilateral phrenic nerve palsy can result in respiratory compromise in an infant. Preoperative ultrasound can help evaluate diaphragm function. Denervation of the intercostals likely results in some long-term chest wall asymmetry that families should be made aware of. Although unlikely, breast development and sensation may also be affected.

Other Nerve Transfers

Other nerve transfers are less common but may be useful in specific circumstances.

The medial pectoral nerve (MPN) may be used as a donor to the axillary or musculocutaneous nerves. Maximal access is gained via an infraclavicular brachial plexus approach. Pectoralis major is retracted or released, and pectoralis minor is released from the coracoid to expose the MPN as it branches from the medial cord. The many branches of MPN can be followed into muscle to attain nerve length. The musculocutaneous nerve is the most lateral branch of the brachial plexus distal to the

coracoid and can be seen travelling through coracobrachialis. The axillary nerve can be found deep to the musculocutaneous nerve.

The use of contralateral C7 (cC7) has significant risks, but may be the only option in the extremely unlikely case of C5–T1 avulsions. Transient donor deficits and persistent synchronous contralateral movements (i.e., donor limb movements when attempting to move the reconstructed limb) are common, and reported results have been variable.

Isolated lower plexus (Klumpke) palsy is extremely rare in infants. In this case, upper plexus donors may be used for lower plexus reinnervation, and brachialis to AIN transfer may be a reasonable option if reconstruction is undertaken.

38.6 Postoperative Care Following Primary Nerve Reconstruction

Infants are immobilized for 3 to 5 weeks. If the shoulder is supple and there is no significant glenoid dysplasia on preoperative MRI, they are secured in a cuff-and-collar sling (► Fig. 38.27a) for 3 weeks, followed by an additional 2 weeks of leaving the arm within the shirt (i.e., arm is not brought through the sleeve) to avoid significant arm motion. If the shoulder is stiff and there is significant GHD, the internal rotators are temporarily denervated with botulinum toxin A and the shoulder is stretched into external rotation using a shoulder spica cast for the full 5 weeks (► Fig. 38.27b). In all cases, range of motion stretching is resumed after 5 weeks.

38.7 Outcomes of Nerve Reconstruction

An in-depth appreciation of outcomes is important for treating providers to manage patients. A comprehensive review of the literature is beyond the scope of this chapter.

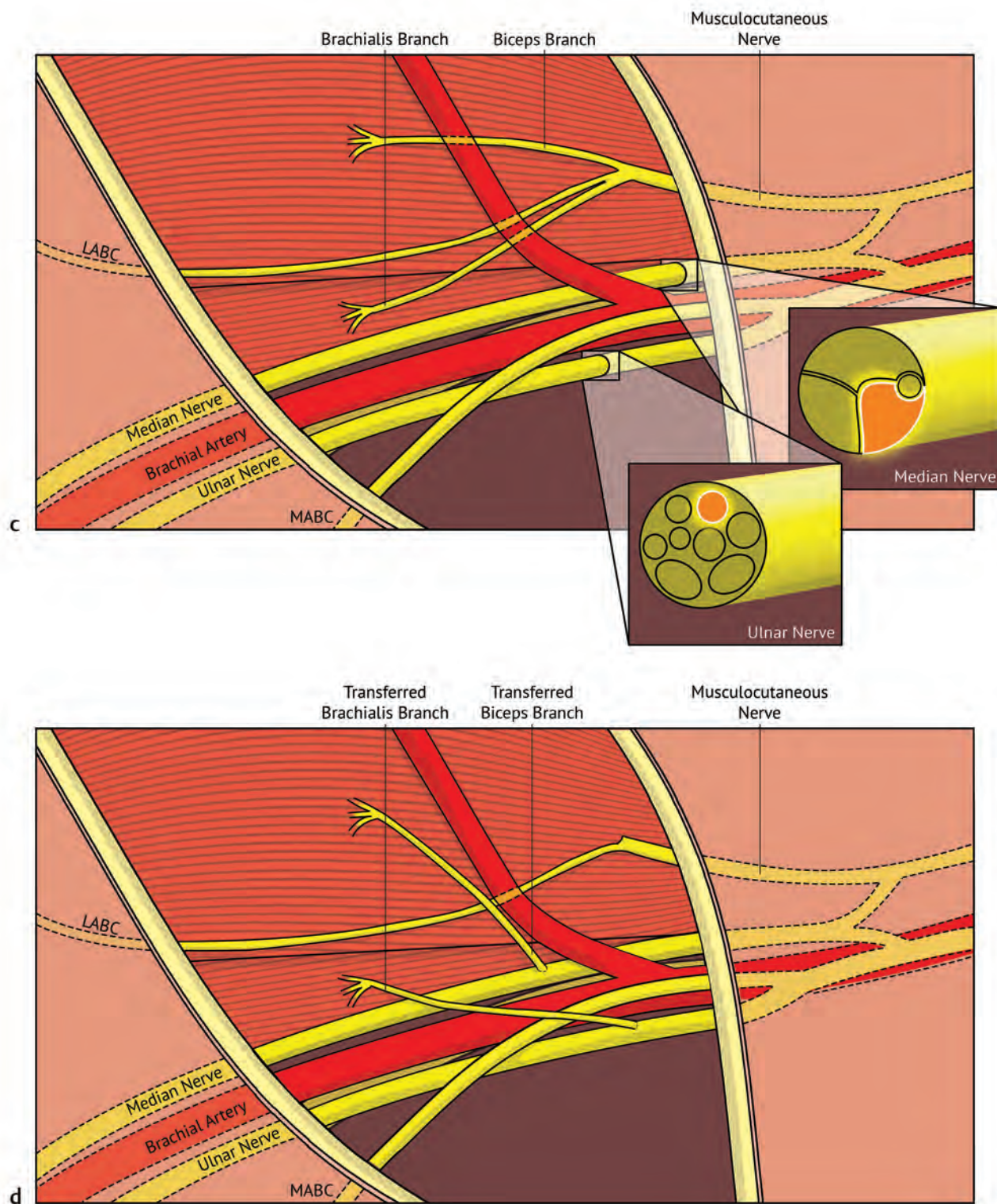


Fig. 38.25 (Continued) (c) Internal topography of median and ulnar nerves. (d) Transfer of median fascicle to biceps branch and ulnar fascicle to brachialis branch.

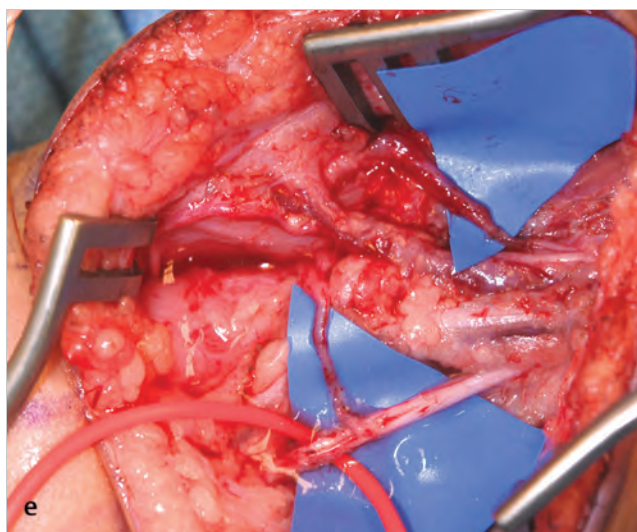


Fig. 38.25 (Continued) (e) Transfer of median fascicle to biceps branch and ulnar fascicle to brachialis branch.

The results of reconstruction using nerve grafts for NBPP have been reported by Lin, Waters, and others (see Lin 2009, Waters 1999, Ashley 2007). The results of reconstruction using nerve transfers for NBPP have been reported by Ladak and Little (see Ladak 2013, Little 2014).

When following the Toronto protocol in the treatment of NBPP, reasonable expectations are for 90% of patients to have improvements, 5 to 10% of patients with no improvements beyond baseline, and 1 to 5% of patients with functional losses related to a complication or failed reinnervation.

Results of reconstruction for TBPP are extremely limited and inferences may be extrapolated from literature on adult traumatic brachial plexus palsies.

Improvements following reconstruction tend to plateau at 2 to 3 years after surgery.

38.8 Secondary Reconstruction

Secondary musculoskeletal reconstruction is generally considered following maximal nerve recovery.

38.8.1 Shoulder

Persistent deficits of external rotation are common in children with brachial plexus palsies, regardless of whether they underwent primary reconstruction or had spontaneous recovery of function. There are several reasons for this. The infraspinatus is the main external rotator of the shoulder, and is innervated by the SSN, which arises from the most cranial fascicular bundle of C5 and the upper trunk. With downward traction, the SSN is susceptible to the greatest forces resulting in injury. Meanwhile, the numerous internal rotators of the shoulder (pectoralis major, subscapularis, teres major, and latissimus dorsi) are innervated by branches originating lower in the plexus and thus their function is often spared. The imbalance of shoulder rotators in the setting of a rapidly growing child is thought to be responsible for progressive contracture and skeletal changes of the shoulder known as GHD.

GHD is characterized by glenoid morphology that progresses from retroversion to loss of the posterior glenoid lip and eventually to a pseudoglenoid. Along with this progression, the humeral head subluxates posteriorly and can take on a flattened morphology (► Fig. 38.28).

Lack of external rotation has significant functional consequences, such as hand to mouth, hand to head, and above shoulder activities. Strategies to address these shoulder deficits can be grouped into those that rebalance the glenohumeral joint and those that salvage function by altering the position of limb range of motion in space.

Reconstruction (Shoulder Release and Tendon Transfer)

Release of shoulder internal rotation contracture can involve capsulotomy, tendon lengthening, ligament release, and osteotomy. There is no consensus on the best method and degree of release to produce the best outcomes, but overly aggressive release can result in scar, morbidity, and loss of internal rotation with significant impacts on overall limb function. In order to avoid excessive release, we start with an open coracohumeral ligament release (► Fig. 38.29a). An anterior shoulder incision provides access to the deltopectoral interval through which the coracoid can be accessed (► Fig. 38.29b). The ligament has fibers that run from medial to lateral, originating from the coracoid. The coracohumeral ligament can be differentiated from the coracoacromial ligament, which has vertical superior course, and the coracobrachialis muscle origin, which has an oblique inferolateral course. Release of the ligament provides space for the humeral head to translate anteriorly with external rotation (► Fig. 38.29c, d), typically allowing an additional 30 degrees of external rotation beyond neutral. If the pectoralis major is tight, the tendinous bony insertion is exposed and a fractional lengthening is performed. With progressive stretch, an additional 30 degrees of external rotation can be gained.

In order to rebalance the rotator cuff, the latissimus dorsi (and sometimes teres major) are transferred to function as external rotators. An incision along the axillary crease provides exposure. The conjoint tendon of latissimus dorsi and teres major is followed to its bony insertion and released (► Fig. 38.29e). Care should be taken to avoid injury to the axillary and radial nerves that course posteriorly along the cephalad and caudal margins of the conjoint tendon, toward the quadrangular and triangular spaces, respectively. The donor muscle is mobilized so that it can insert into the greater tuberosity of the humerus (► Fig. 38.29f). Splitting the interval between triceps and deltoid exposes the new site of insertion. If dissecting too distal, the sensory branch of axillary nerve and associated blood vessels may be encountered. The axillary nerve is in the vicinity and should be preserved.

The shoulder is immobilized in external rotation using a shoulder spica cast for 4 weeks. Therapy and stretching are initiated at 4 weeks, and strengthening is initiated at 8 weeks after surgery. Glenohumeral remodeling and better clinical results have been demonstrated with shoulder release and tendon transfer when children undergo surgery at 2 to 4 years of age. Glenoid osteotomy to correct glenoid version has been described as an adjunct and may allow for improved motion in

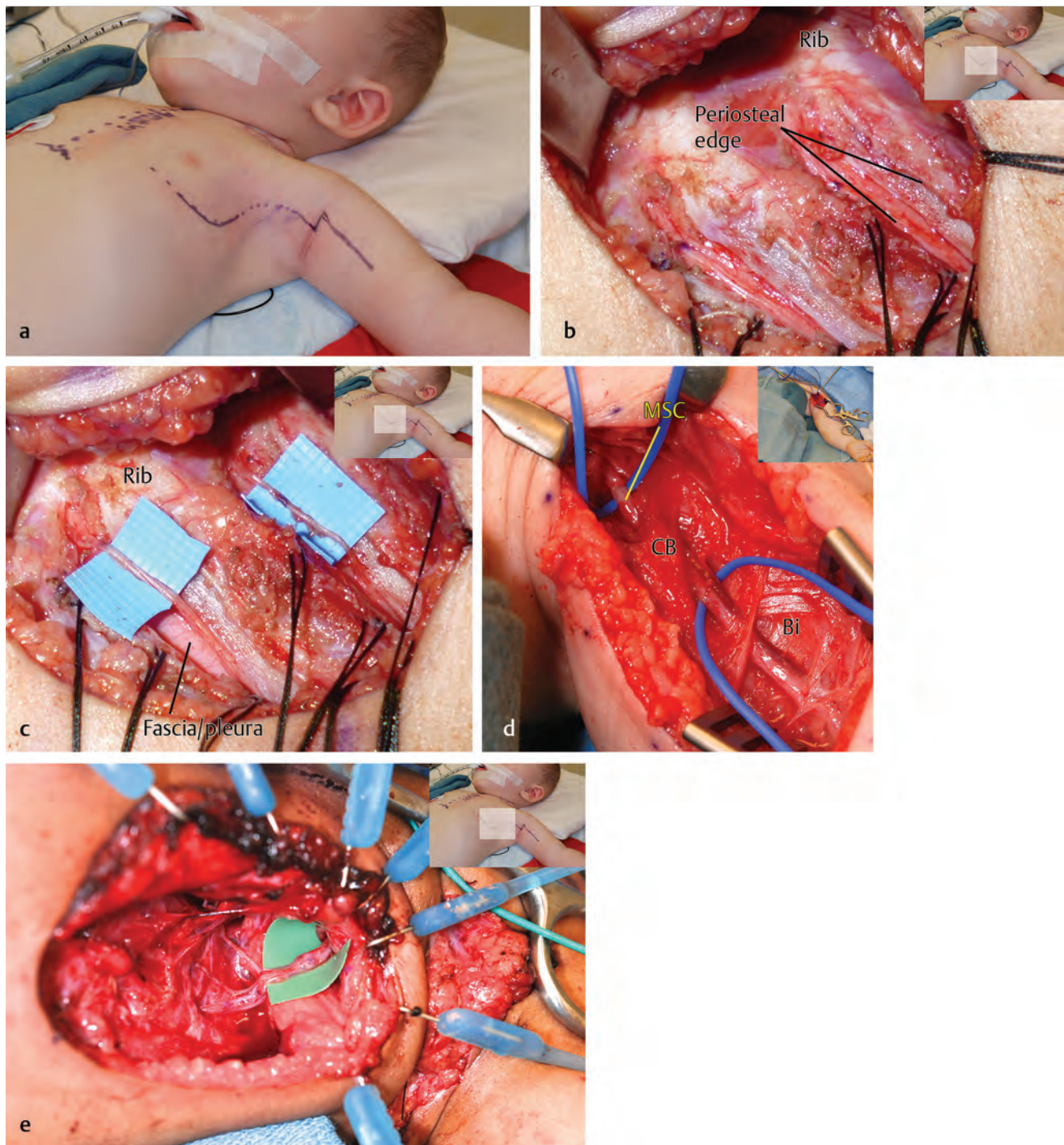


Fig. 38.26 Intercostal to musculocutaneous nerve transfer (ICN-MSC). (a) Anticipated inframammary fold and planned incisions. (b) Retraction of pectoralis major and exposure of chest wall. Periosteum on the anterior surface of the rib has been incised and elevated off the lower border and posterior surface of the bone. (c) Sutures along periosteal edge allow downward traction to bring intercostal neurovascular structures out from the undersurface of the ribs. Incision through periosteum allows direct exposure of nerve. (d) Medial upper arm incision allows exposure of musculocutaneous nerve (MSC) as it courses through the coracobrachialis muscle (CB). (e) The nerve is followed up to lateral cord E. Musculocutaneous nerve has been passed through axilla and intercostal nerves have been tunneled through serratus anterior to allow direct coaptation.



Fig. 38.27 Postoperative immobilization. (a) Cuff and collar sling. (b) Shoulder spica cast. External rotation reduces the humeral head anteriorly into the glenoid fossa.

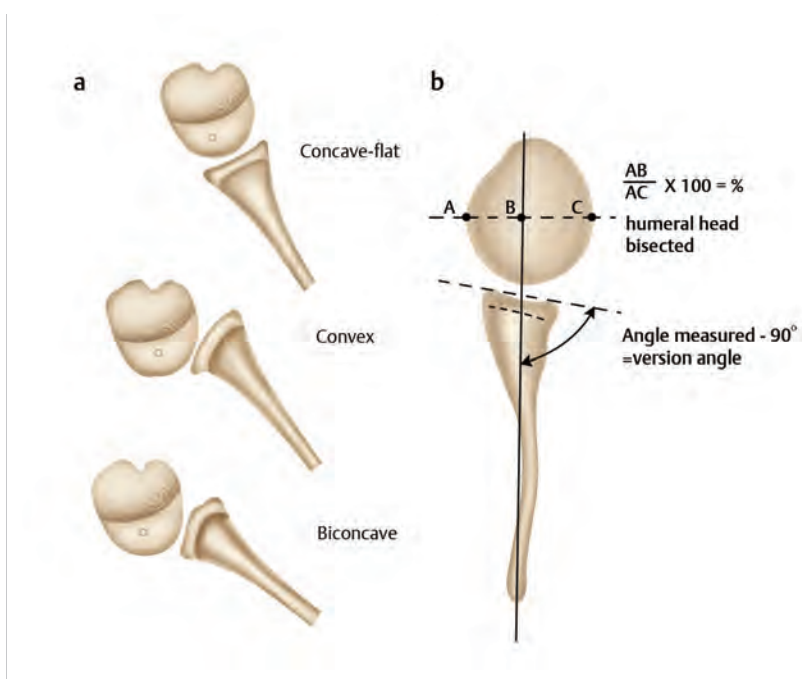


Fig. 38.28 (a) Progressive glenoid dysplasia: concave-flat, convex, biconcave. (b) Measurement of glenoid version and posterior subluxation according to the percentage of humeral head anterior to axis of scapula.

older children with less potential for glenohumeral remodeling; however, long-term outcomes and risks remain unknown.

Salvage (Humerus Osteotomy)

In the case of severe GHD that is unlikely to remodel, or if further glenohumeral reconstruction is unlikely to provide functional improvement, humeral rotation osteotomy may be an option to reposition the available active shoulder rotation into a more functional position in space. The available range of

motion should be assessed and the planned shift in rotation should be measured and agreed upon with the patient before surgery. The humeral shaft is cut and rotation to the new position is stabilized with rigid fixation (► Fig. 38.30).

38.8.2 Elbow

Failure to recover elbow flexion following nerve reconstruction is rare. In the case of inadequate or failed recovery, several options to augment or produce elbow flexion have been

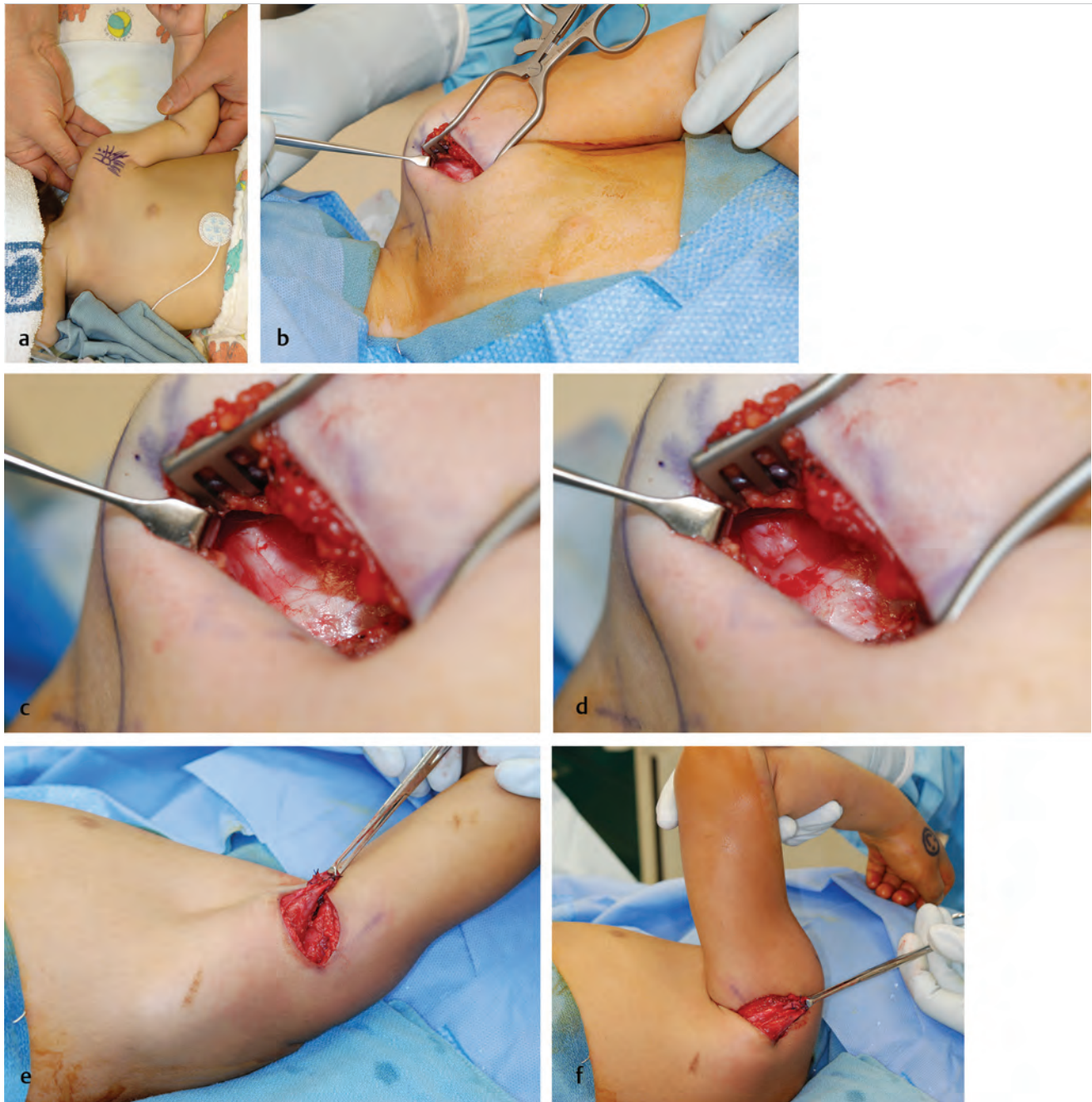


Fig. 38.29 Shoulder release and tendon transfer. (a) Exposure of coracohumeral ligament via anterior incision. Dissection through deltopectoral interval exposes coracoid. Dissection lateral to coracoid reveals ligament. (b) Shoulder release and tendon transfer. Coracohumeral ligament blocks external rotation. (c) Shoulder release and tendon transfer. The humeral head protrudes with external rotation following coracohumeral ligament release. Additional release can be attained by fractional lengthening of pectoralis major through the same incision. (d) Shoulder release and tendon transfer. A separate axillary incision provides exposure to the conjoint tendon of latissimus dorsi and teres major. The tendon is released from humerus taking a small cuff of periosteum. (e) Shoulder release and tendon transfer. (f) The tendon is passed through the interval between triceps and deltoid to insert into the greater tubercle.

described. An individualized plan should be devised. If elbow flexion is weak and the forearm flexor pronator group is strong, the flexor pronator group origin can be translocated proximally so that this muscle group augments elbow flexion (Steindler transfer). Bipolar transfer of the latissimus dorsi or pectoralis major to reconstruct elbow flexion has been described; however, these motors may not be fully expendable in an already compromised limb.

Free muscle transfer is an option that adds an additional motor to the limb, without sacrificing any limb function. A free gracilis muscle can be transferred and innervated with intercostal or other donor nerves. The distal tendon of the gracilis can be harvested, allowing the tendon to be woven into the native biceps insertion (► Fig. 38.31). Because the gracilis is longer than the native elbow flexors, it must be tensioned appropriately by shortening the proximal origin or by fixing the origin superior to the level of the coracoid (i.e., wrapping around lateral clavicle).

38.9 Musculoskeletal Considerations and Complications

Management of skeletal changes resulting from chronic denervation in a growing child has traditionally been postponed until the child is older than 2 years. However, posterior shoulder subluxation and dislocation have been observed in infants. A recent review of MRI performed prior to nerve reconstruction revealed an extremely high incidence of significant GHD (74%) in spite of nonsurgical treatments and therapy. These skeletal changes can be seen as early as 3 months of age. Loss of passive range of motion in external rotation is a significant clinical finding. A high axillary crease or additional upper limb skin creases should be noted (► Fig. 38.32) as these can result from limb foreshortening with posterior subluxation.

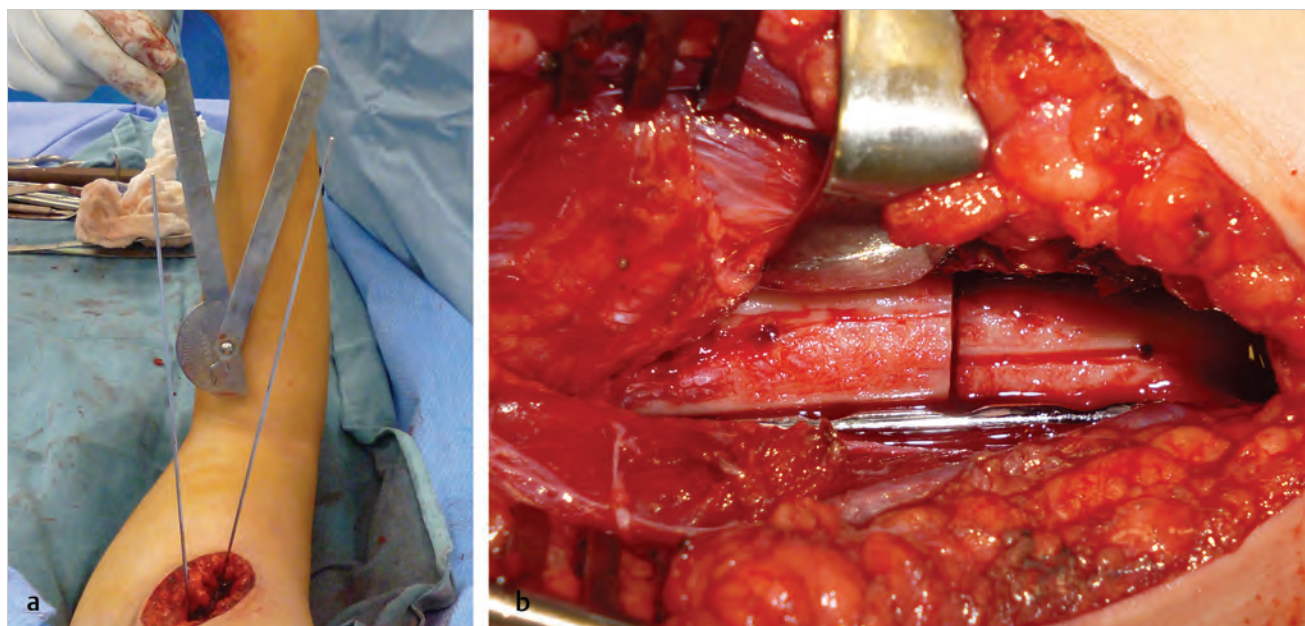


Fig. 38.30 Humerus rotation osteotomy. (a) The medial arm exposure is seen from above the patient's head. K-wires are used to measure appropriate rotation following transverse osteotomy. (b) Humerus rotation osteotomy. Plate fixation following osteotomy and rotation. A longitudinal slot was made using a burr for orientation. Note displacement following rotation. K-wires for orientation have been removed.

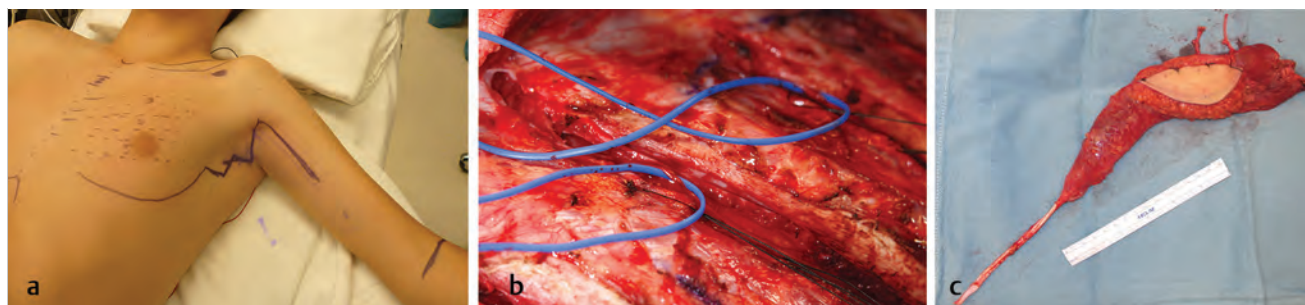


Fig. 38.31 Free gracilis transfer for elbow flexion. (a) Planned incisions for intercostal nerve harvest and flap inset, including monitoring skin paddle. (b) Intercostal nerve harvest. (c) Gracilis muscle harvested with obturator nerve for re-innervation and long distal tendon to weave into biceps tendon.



Fig. 38.32 Arm and axillary creases. An additional upper arm crease and a deep axillary crease that extends superiorly on the baby's right are due to bony foreshortening from glenohumeral subluxation.

Given the impact of shoulder dysplasia on ultimate functional outcomes, our center now performs routine shoulder ultrasounds at 3 and 6 months of age in all infants with persistent motor deficits. An alpha angle greater than 30 degrees is considered abnormal (► Fig. 38.33). If there is no loss of passive range of motion, we use botulinum toxin A to weaken the internal rotators, which allows families to continue shoulder stretching. When there is an associated loss of passive range of motion, suspected dislocation, or dysplasia of the posterior lip of the glenoid, chemodenervation is combined with shoulder spica casting in external rotation for 6 weeks. External shoulder rotation reduces the humeral head in the glenoid fossa, thereby encouraging glenoid remodeling around a centrally reduced humeral head (► Fig. 38.34).

There have been recent reports that early infant shoulder repositioning may help avoid shoulder subluxation and encourage motor recovery. We have adopted the Super splint described by Verchere et al, in situations where families can return for frequent follow-up. The splint positions the forearm into supination and the shoulder into external rotation. Other centers are also accumulating experience with this strategy.

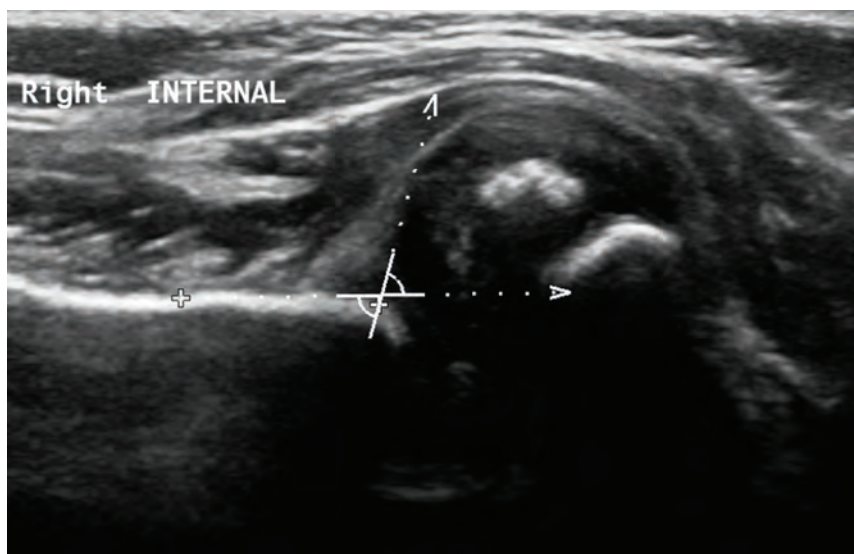


Fig. 38.33 Shoulder ultrasound with severe subluxation. Lack of ossification makes the infant shoulder difficult to assess on X-ray. Ultrasound allows visualization of the humeral head and its relationship to the glenoid. In this view, the posterior border of scapula can be seen. A line from the posterior labrum that is tangent to the humeral head forms an angle as it intersects with the posterior border of scapula. This angle should be 30 degrees or less.

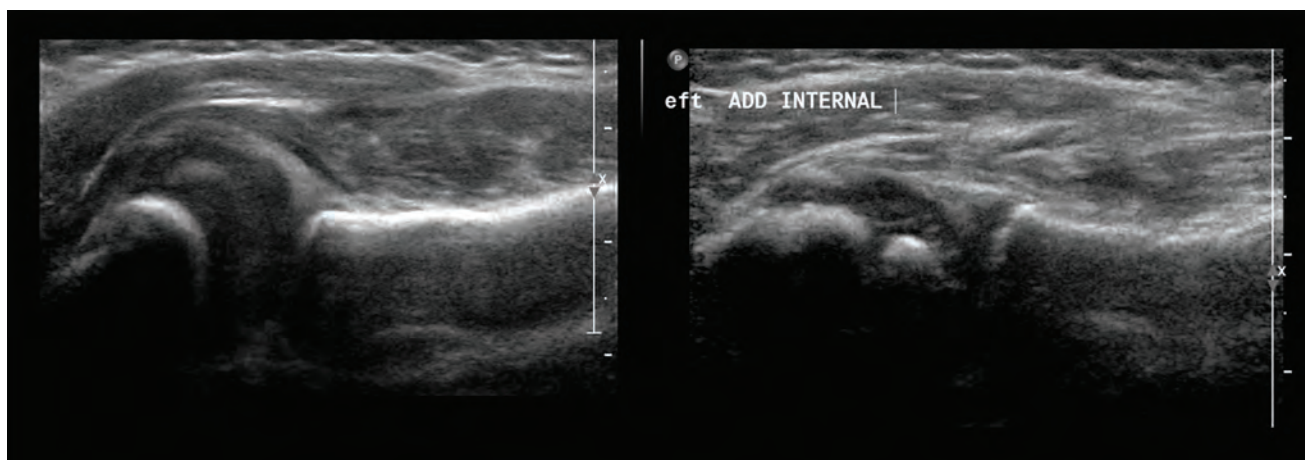


Fig. 38.34 Changes with chemodenervation and casting. Pretreatment on the left. Posttreatment on the right with relocation of glenohumeral joint and posterior labrum.

38.10 Key Points

- The complexity of pediatric brachial plexus palsy and the benefits of multidisciplinary care warrant treatment in specialized centers that can provide the breadth of team care.
- Given the musculoskeletal and neurodevelopmental consequences of chronic denervation, nonsurgical treatment is critical in the outcome of pediatric brachial plexus palsy, regardless of whether nerve and/or musculoskeletal reconstruction is undertaken.
- Longitudinal assessment to understand the trajectory of pediatric brachial plexus palsy is necessary in deciding upon nerve reconstruction.
- The role of nerve transfers as the primary strategy in reconstruction of NBPP is unclear. Nerve transfers alone are inadequate for reconstruction of palsies more severe than Erb palsy (type 1 palsy) and nerve grafts should be used in those situations.
- Nerve transfers are useful in the case of inadequate proximal axons (i.e., avulsions), isolated deficits, late presentation, and failed primary reconstruction.

Suggested Readings

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39 Pediatric Hand Trauma

Amir H. Taghinia

Summary

Hand trauma in children typically results from sports and play. Compared to adults, children experience less stiffness and better nerve regeneration. Immobilization is often important following operative intervention to ensure the most favorable outcome.

Keywords: pediatric, trauma, hand, fracture, tendon, nerve

39.1 Introduction

The surgeon's approach to hand trauma in children should be different from hand trauma in adults. The mechanism of injury, anatomy, healing, and psychosocial concerns of a child require special attention. As opposed to adults, injuries in children vary substantially by age. Young children usually present with "injuries of curiosity"—such as those inflicted by handling broken glass or inserting a finger in the spokes of a spinning bicycle wheel. Older children and adolescents sustain hand injuries from sports and play. Luckily, large industrial hand injuries are rarely seen in children.

Children are usually free of significant medical comorbidities, and thus heal better and faster. Stiffness is unusual, so it is easier for the surgeon to balance healing with commencement of activities. Children regenerate nerves better and rarely develop painful neuromata or pain syndromes. However, in children the important hand structures are small and there is little margin for technical error. Diagnosis can be difficult in a young child or uncooperative adolescent. The surgeon has to respect growth and change over time. In more severe injuries, the surgeon may need to spend significant time counseling the parents and dealing with psychosocial issues. Lastly, there are a number of unusual injury variants in children that if misdiagnosed or mistreated can lead to substantial morbidity.

Adequate immobilization is crucial for treating children with hand injuries. Pain usually subsides several days after injury and children are likely to resume their usual activities if they are not held back. Inadequate immobilization is usually the main culprit in malunions, dehiscent wounds, and tendon ruptures. Because stiffness is rarely a problem, there is very little downside to adequate immobilization.

39.2 Diagnosis

Observation, physical examination, and plain radiography are the main diagnostic tools. In an uncooperative, anxious, or hysterical child, observation is sometimes the most important factor. The importance of distal examination cannot be overemphasized. Exploring a child's hand or forearm in the emergency department to ascertain for presence of injury is difficult and dangerous. A more gentle approach to examination, such as by recruiting the help of a child life specialist or touch pad game to distract the child, is usually much more effective than brute force. The most difficult injuries to diagnose are flexor tendon

and digital nerve injuries in infants and young toddlers, even 5 to 7 days postinjury. Repairing an old flexor tendon injury in a child is very difficult, so exploration is warranted if there is any suspicion.

Plain radiographs usually detect most fractures and dislocations, with the notable exception of acute nondisplaced scaphoid fractures. Ultrasound is instrumental in detecting vascular injury, or more commonly, foreign bodies—especially wood. Computed tomography or magnetic resonance imaging is rarely needed.

39.3 Fractures

In the skeletally immature patient, the ligaments are stronger than bones. Accordingly, it is rare to see ligamentous injury and more common to see fractures. Fractures occur in weak areas of the metaphysis, physis (growth plate), and epiphysis, and result in predictable injury patterns, outlined in the Salter–Harris classification. The most common hand fracture is a proximal phalangeal fracture with a Salter–Harris type II configuration, usually caused by a deviating force (► Fig. 39.1).

Most pediatric hand fractures are nondisplaced or minimally displaced and only require immobilization for 2 to 3 weeks, followed by slow return to normal activity. Most displaced fractures can be reduced in the emergency room (ER) or the operating room under local block, sedation, or general anesthesia. Unstable fractures are usually treated with closed reduction and percutaneous pinning. An open approach is rarely needed.

Fracture remodeling in children can and does occur (► Fig. 39.2). The closer a fracture is to the physis and the younger the patient, the higher is the remodeling potential of the fracture. However, caution should be exercised in treating all fractures with the hopes that they will remodel. Severely displaced fractures will not completely remodel, nor will those far away from the physis (e.g., displaced phalangeal condylar neck fractures). As a general rule, deviation of a finger *in* the plane of motion is tolerated, whereas deviation *perpendicular* to the plane of motion is not.

Several pediatric fracture variants can be tricky to diagnose or treat; these are outlined below.

39.3.1 Ulnar Collateral Ligament Injury Variant

This injury is analogous to an adult ulnar collateral ligament injury except that the epiphysis breaks, resulting in a Salter–Harris type III injury (► Fig. 39.3). It occurs from a radially directed force in adolescents near skeletal maturity. The ligament is attached to the small bone fragment. An open approach is required through a dorsal incision. The adductor aponeurosis is incised and a dorsal arthrotomy exposes the joint. The bone is reduced with careful re-approximation of the articular surface. One or two 0.035-in wires can be used to fixate the bone. The bone fragment is small, so it is best to get the fixation as good as possible before inserting the pins. A thumb spica cast is

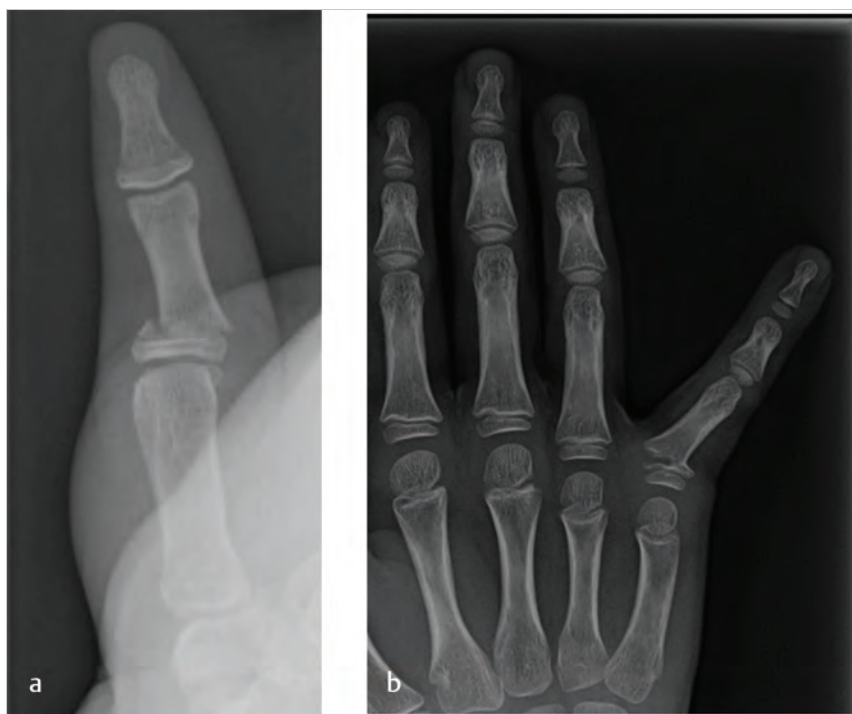


Fig. 39.1 Examples of displaced Salter–Harris type II fractures of the proximal phalanges (**a**) in the thumb and (**b**) in the small finger. The small finger proximal phalangeal fracture is best reduced by flexing the metacarpophalangeal (MP) joint to 90 degrees, placing a pen in the fourth webspace, and applying radially directed digital pressure on the distal aspect of the proximal phalanx. MP joint flexion tightens the collateral ligaments and prevents motion at the MP joint. The pen acts as a fulcrum upon which the proximal phalanx rotates.

applied for 4 weeks. The pins are then removed, gentle therapy commences, and the thumb is protected in a splint for another 2 to 3 weeks.

39.3.2 Phalangeal Condylar Neck Fractures

Condylar neck fractures in children are the result of hyperextension injuries. They occur in the proximal or middle phalanges. The condyle is attached to the distal bone via collateral ligaments and the volar plate. The bone usually fractures transversely, but the ligaments remain attached. The condylar piece will then displace and angulate dorsally. If allowed to heal in this configuration, the retrocondylar recess is ablated and prevents full flexion of the joint. Closed reduction, pinning, and cast immobilization are advocated. If the fracture appears well reduced initially, it should be immobilized (with a cast) and watched closely with serial radiographs as it can displace. It is not uncommon for these children to present several weeks postinjury with an impending malunion. Waters presented an effective technique of percutaneous osteoclasis (using a pin) and reduction. This technique avoids an open approach that can compromise the tenuous blood supply of the distal condylar segment.

39.3.3 Seymour Fracture

This injury is a distal phalangeal juxtaaphyseal injury that results from a sudden flexion force applied to an extended distal phalanx (► Fig. 39.4). It is analogous to an adult mallet injury except the injury occurs at the growth plate. The extensor attaches to the dorsal epiphysis, but the attachment of the flexor tendon is broader—onto the epiphysis as well as the distal fracture fragment. Accordingly, there is a flexion moment on the distal fragment. A severe injury will displace the distal

fragment dorsally lacerating the nail bed—usually at the depth of the recess where the germinal matrix meets the eponychial fold. The bone/nail bed complex then displaces the nail fold and herniates dorsally, thereafter sitting on top of the nail fold (► Fig. 39.5). This is an open fracture and has to be recognized as such. There is much misrepresentation in the literature and standardized exams about the nature of this injury. It is the nail *fold*, not the nail *bed*, that gets trapped in the fracture and disallows easy reduction. In the case that is demonstrated (► Fig. 39.5), one can see the entire nail plate—it is totally adherent to the nail bed and there is no excess nail bed trapped in the fracture. Treatment requires reduction of the fracture (usually with relaxing incisions in the nail fold) and pin fixation. The fracture heals quickly. This injury is misdiagnosed often, and thus treatment can be delayed, resulting in chronic nonunion and significant nail abnormalities.

39.4 Joint Injuries

Sprains of the proximal interphalangeal joint and metacarpophalangeal joint are common, especially in young athletes. These usually occur with a small chip fracture. A short period of immobilization is usually necessary to allow the swelling and pain to subside. Most patients can start range of motion exercises within 2 weeks and then return to normal activities within a month.

Joint dislocations in infants and toddlers are extremely rare. Irreducible trigger thumbs are commonly misdiagnosed as dislocations in the toddlers. Dislocations are more common in older children and adults. The most common joint is the finger proximal interphalangeal joint. Reduction in children is similar to that in adults.

Thumb metacarpophalangeal joint dislocation occurs commonly in older children. It is usually the result of a fall in the 6

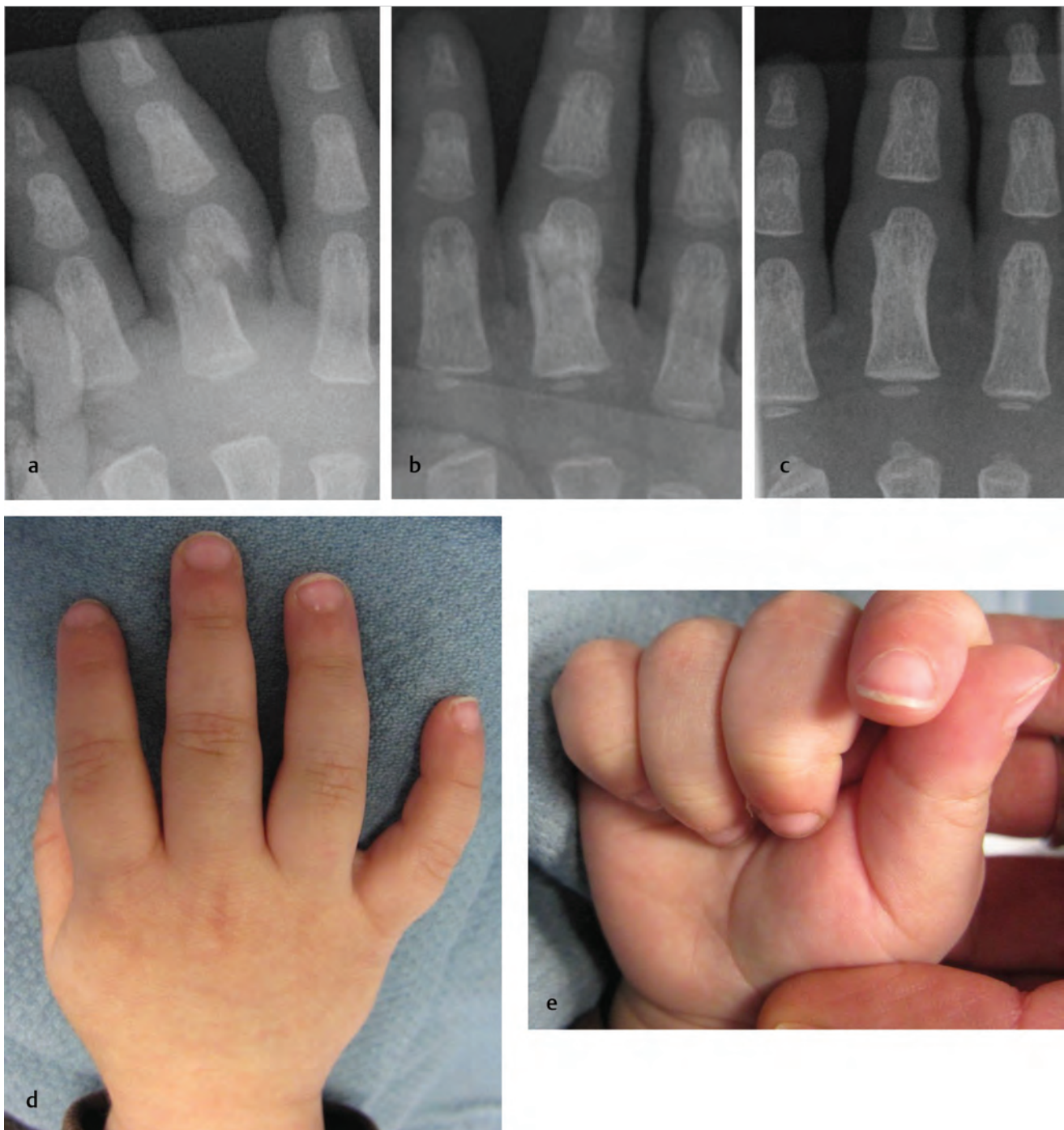


Fig. 39.2 Displaced proximal phalangeal fracture. No intervention was undertaken on this (a) acute fracture. (b) After 1 month, there is mild remodeling. (c) After 3 months, the remodeling continues to improve the alignment and appearance. (d) and (e) demonstrate excellent function and range of motion at 3 months post injury.

to 10 age group. Reducing these injuries can be tricky, so sedation and a wrist block are necessary adjuncts. The reduction maneuver is more about finesse than force. The surgeon places the dominant thumb pad on the dorsal base of the proximal phalanx and stabilizes the metacarpal on the palmar side with the dominant fingers. The other hand acts as an assistant as a gentle pushing motion of the thumb and simultaneous pulling motion of the fingers roll the head of the proximal phalanx onto

the metacarpal head. A few attempts may be needed. One should not distract the thumb distally as this prevents reduction, instead of helping it.

39.5 Tendon Injuries

Flexor tendon injuries can be difficult to diagnose in toddlers. If there is any doubt, early exploration is recommended as

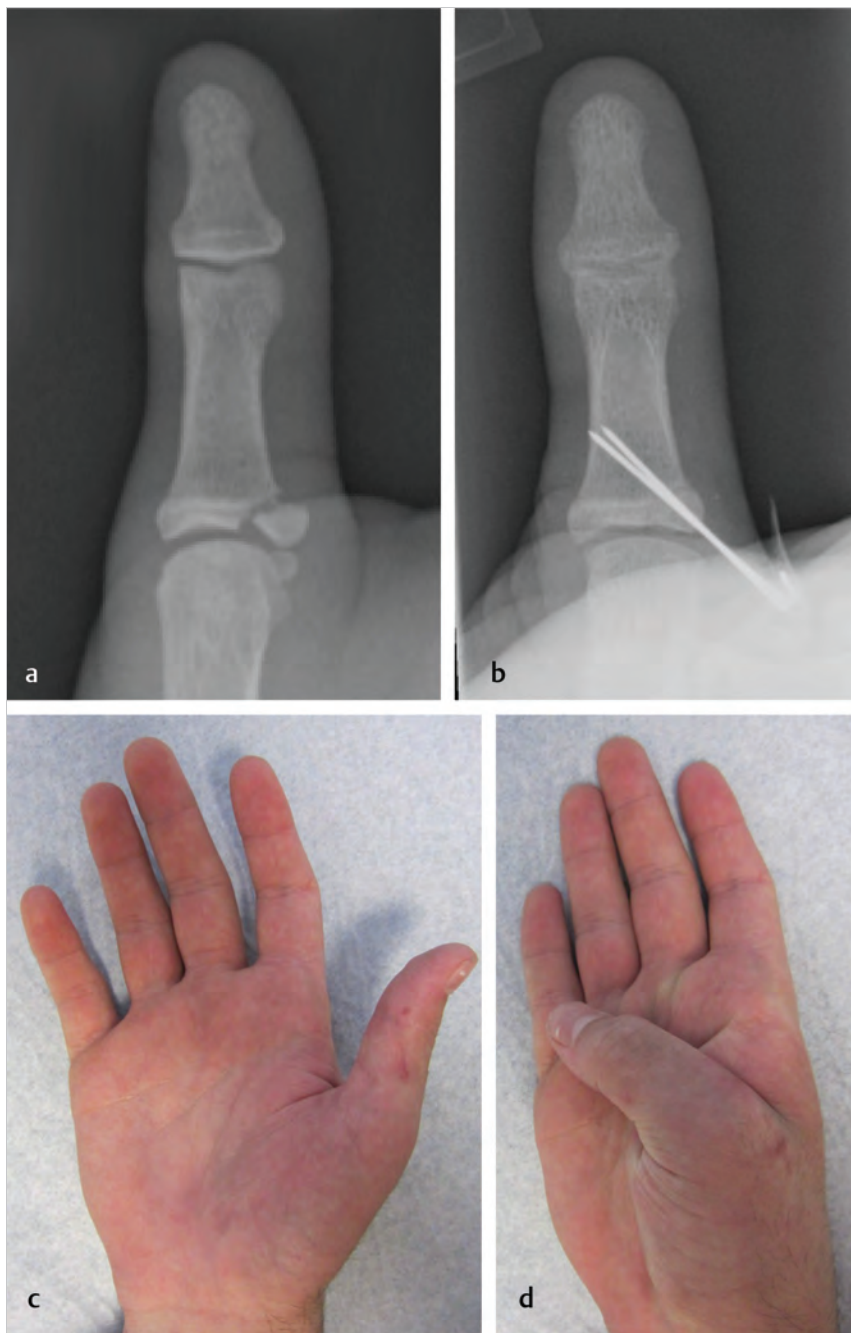


Fig. 39.3 Thumb metacarpophalangeal joint ulnar collateral ligament variant injury in adolescents. Instead of avulsing the ligament, adolescents sustain (a) Salter–Harris type III fractures where the ligament remains attached to the epiphyseal fragment. (b) This injury should be treated via an open approach and direct visualization and reduction of the fracture. (c,d) Three months postoperatively, this patient demonstrates excellent function.

delayed repair can be quite difficult. The technical execution of repairing a flexor tendon is challenging as the structures are small and maneuvering around the pulleys and avoiding bulky repairs is difficult. Primary repair is advocated for acute injuries. A four-core repair with epitendinous suture is usually sufficient (► Fig. 39.6).

The choice of postoperative rehabilitation protocol should be based on the compliance of the patient. Because joint stiffness is rare, the surgeon can err toward longer immobilization to protect the repair. Older teenagers can usually comply with adult flexor tendon rehabilitation protocols. Younger teens and children should be immobilized in a cast for 3 to 4 weeks. After that, a modified, more aggressive flexor tendon protocol can

begin. A similar approach is taken for most extensor tendon injuries of the fingers and the hand.

Most authors recommend against staged flexor tendon reconstruction in children, mainly due to compliance issues. However, staged reconstruction is possible and can be successful in the right patient—careful patient selection is critical.

39.6 Nerve Injuries

Functional return after nerve injuries in children is much better than in adults. There are several explanations for this difference. The distances to regenerate are smaller in children, so nerves can reach their distal targets more quickly. Axonal

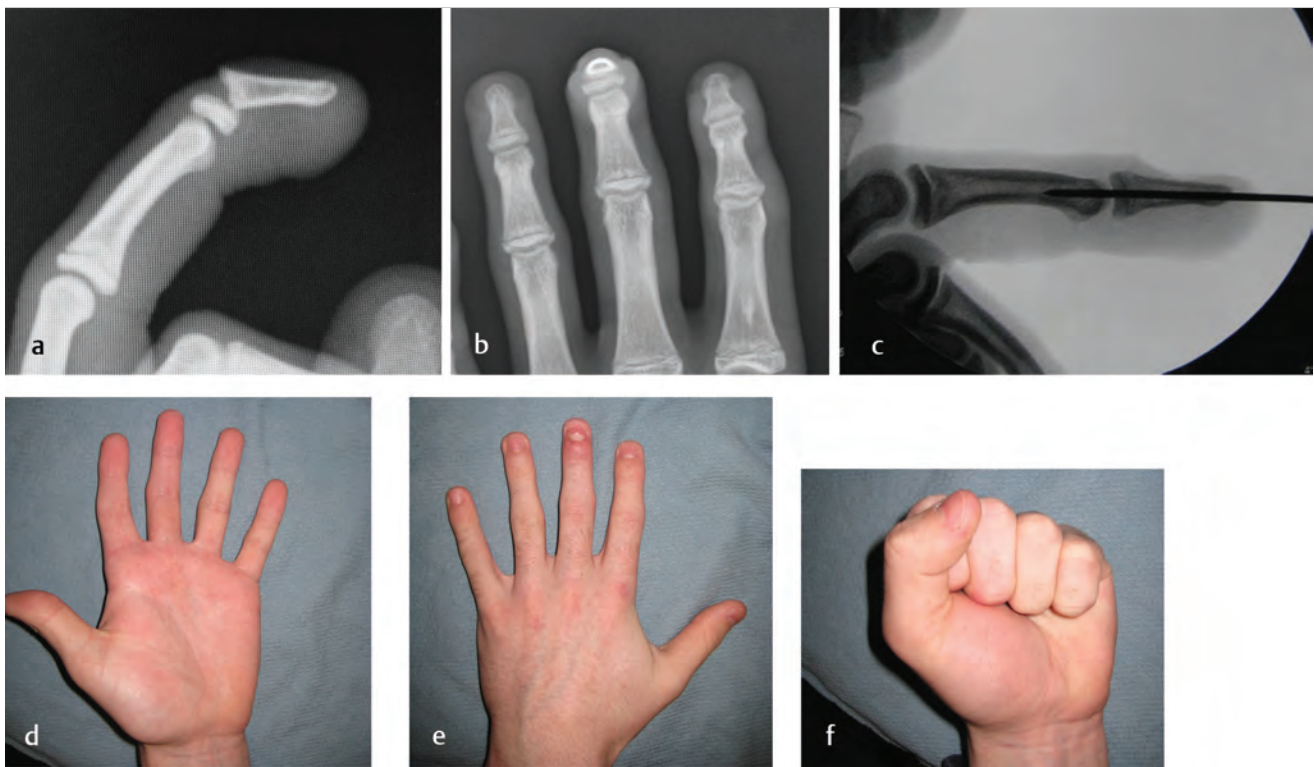


Fig. 39.4 Seymour fracture. The radiographs in (a) and (b) demonstrate a juxtaphyseal fracture which herniates above the nail fold. The nail fold prevents reduction. Lateral incisions in the nail fold (made perpendicular to the nail fold) are usually required to reduce the fracture and a pin secures it (c). Postoperative result in the same patient 2 months postoperatively (d–f) demonstrates normal active range of motion.



Fig. 39.5 Acute Seymour fracture. The clinical views in (a) and (b) are typical of a displaced fracture. The bone distal to the growth plate remains attached to the entire nail plate and nail bed. The nail bed remains completely intact, with the entire attached nail plate. The area of injury/laceration is at the depth of the recess where the nail bed meets the nail fold. Once reduced, this area cannot actually be repaired as it lies in a recess. The author's experience is that the nail plate does not need to be removed, nor does the nail bed require repair. Reduction (usually by the help of relaxing incisions in the nail fold) and pin fixation are all that is needed.

sprouting in children is more robust. And finally, children have better cortical plasticity, so the brain adapts to rewiring mismatches more easily (► Fig. 39.7).

39.7 Fingertip Injuries

Fingertip injuries are very common in younger children. The most common mechanism of injury is a crush injury, usually

caused by a door. The degree of injury is variable and depends on the force. It can range from simple subungual hematoma to an open fracture with nail bed laceration to amputation (► Fig. 39.8). As the nail gets crushed at the level of the sterile matrix, its proximal end gets lifted off with a small bit of eponychium. The nail bed develops a laceration with or without a tuft fracture. Repair involves reconstructing the nail bed and placing the old nail or a piece of petroleum gauze as a

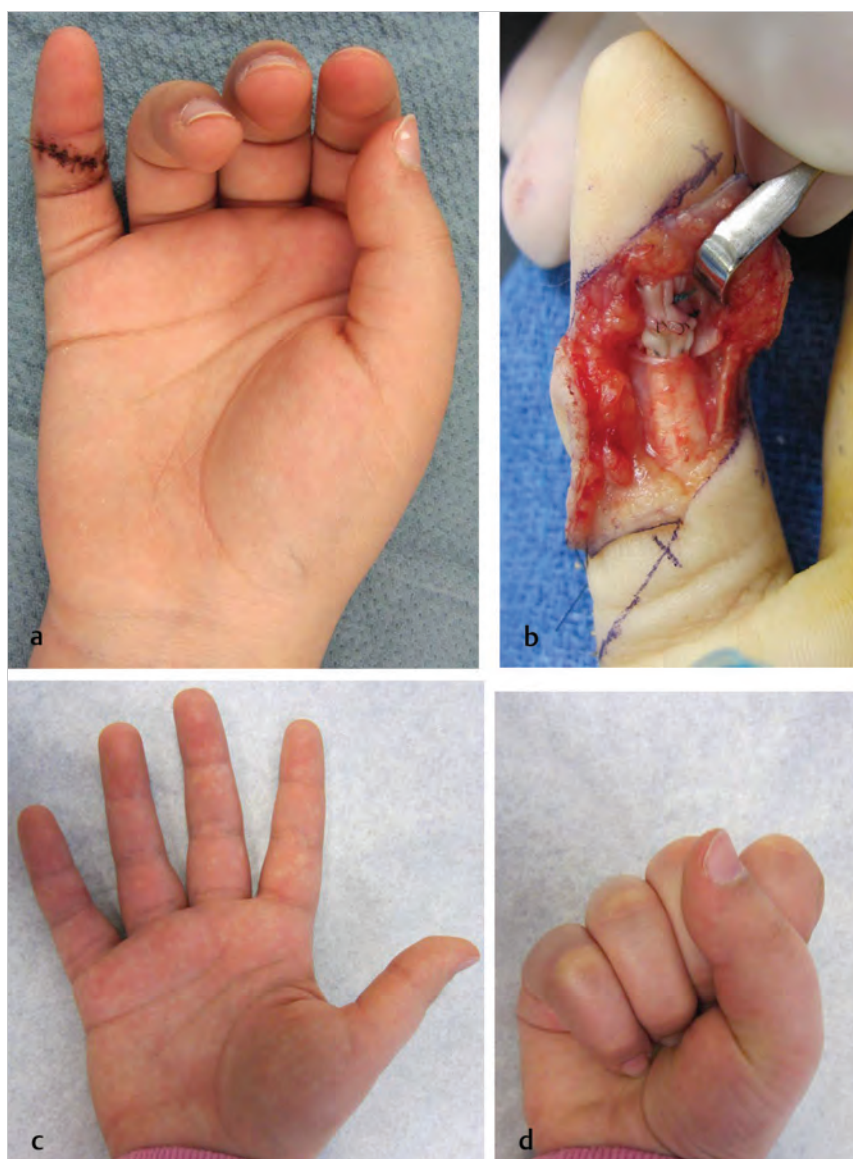


Fig. 39.6 Zone II flexor tendon repair in a 12-year-old female. (a) The flexor digitorum profundus tendon alone was lacerated. (b) A four-core repair followed by epitendinous suture is performed. (c,d) Postoperative result 3 months later demonstrates normal active range of motion.

stent under the nail fold. The tuft fractures rarely need intervention.

In more severe injuries, the distal skin and nail bed can be amputated. The common teaching is to replace the amputated tip as a composite graft and that it usually takes well in small children. We studied this in our institution in a large series of patients and found that only 40% took fully. Nevertheless, most patients healed with minor deformity and no functional consequence without the need for any additional procedures. At best, the graft takes and heals well; at worst, it provides a biological dressing that will ultimately slough as the skin grows under it.

39.8 Amputations

An amputation injury in a child is uniformly devastating for parents. The loss creates feelings of guilt, sorrow, and remorse that overwhelm most parents. Most will “do anything” including giving up their own limbs for their child to be whole again.

The surgeon treating these patients often spends more time counseling the parents and children than providing direct clinical care, especially in more proximal injuries where reconstruction or replantation is not possible. In the case where replantation is not possible or not successful, the surgeon should consider revision amputation a reconstructive procedure, not a failure.

In treating patients with amputations, the primary goal is to maximize functional length. If there is extra bone, then soft-tissue coverage in the form of local or free tissue transfer should be considered. One should preserve growth plates and joints if possible. About 80% of the growth of the radius is owed to the distal radial growth plate, as compared to 20% for the proximal radial physis. In children, metaphyseal or diaphyseal amputation stumps can overgrow irregularly, creating heterotopic ossification and sharp spicules that perforate the skin and are painful. Preserving the entire bony length is thus important to avoid subsequent complications related to growth.

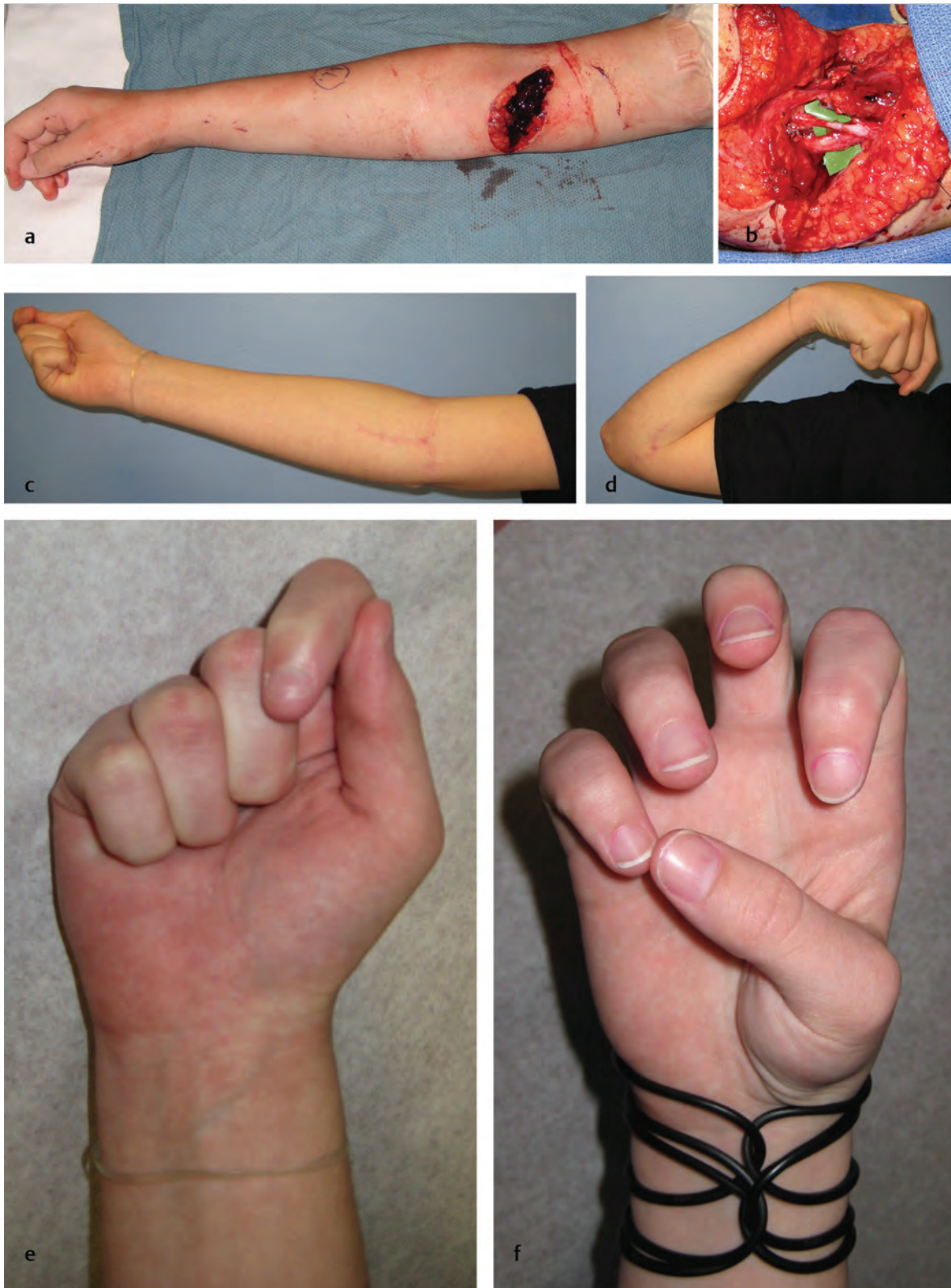


Fig. 39.7 Median nerve injury in a 14-year-old female. (a) This patient sustained a glass injury to the dominant antecubital fossa, resulting in laceration of the brachial artery, median nerve, and brachialis muscle. (b) These structures were repaired on the day of injury. (c,d) Three months postoperatively, she had regained elbow and digital flexion, though she had not recovered motor function of the anterior interosseous nerve. (e) She was unable to flex the index finger distal interphalangeal and the thumb interphalangeal joints. (f) Several months later, the acute interstitial nephritis recovered fully as did intrinsic muscles of the thumb.



Fig. 39.8 Crush injury with fingertip amputation. (a,b) This 16-year-old female sustained a crush injury with exposed distal phalanx. (c,d) A thenar flap was used to cover the bone and avoid shortening. (e,f) The flap was divided 2 weeks later and allowed to heal spontaneously without any inset. Three months later, the patient had a well-contoured stump with normal range of motion of the proximal interphalangeal joint.

39.9 Replantation

Children rarely develop stiffness or chronic nerve-related pain issues. The concept of speeding “return to work” does not apply, and children have healthy vessels and robust nerve regeneration. Accordingly, the pediatric hand surgeon can be more aggressive in attempting replantation. The author’s experience

is that digital replantations are—on the whole—much easier in children than in adults. Dissecting the tissues and finding the important structures is easier and faster. The vessels can undergo significant stretch without causing undue tension. Subsequent recovery is smoother—as rarely does one see bony nonunion, soft-tissue atrophy, pain syndromes, or other complications of adult replantation. The main challenge is technical

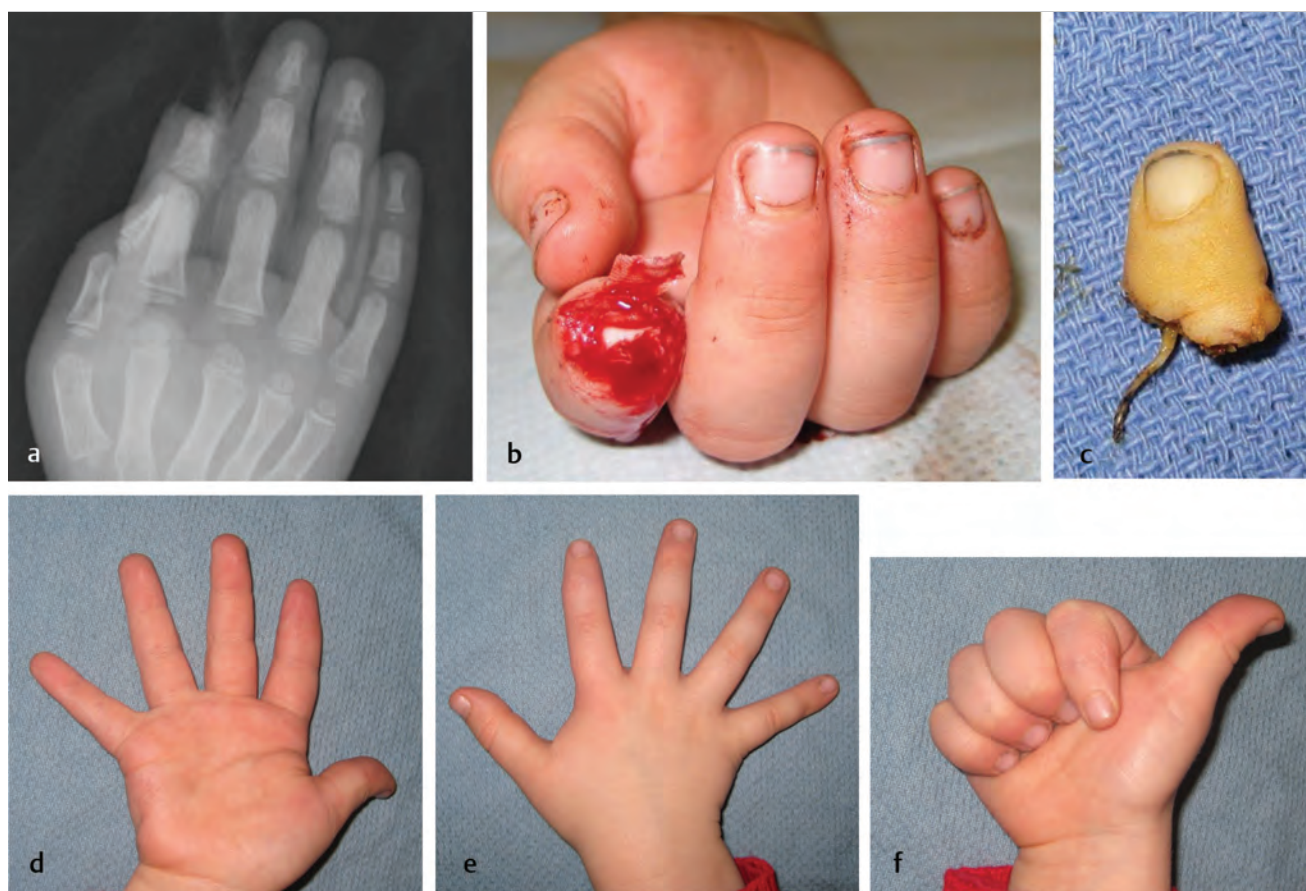


Fig. 39.9 Replantation. (a) This 3-year-old male sustained a complete amputation of the dominant index finger at the distal interphalangeal (DIP) joint. (b) The cartilage of the middle phalangeal condyle was missing and (c) the radial digital nerve was avulsed. The digit was replanted; one digital artery and two digital veins were repaired. The finger survived. (d-f) One year postoperatively, the patient has very little motion at the DIP joint. Four years postoperatively, the patient has normal sensation with no cold intolerance or pain symptoms, but the DIP remains stiff.

execution of repairing the vessels, especially small dorsal veins, which must be done with an operating microscope and ultra-fine instruments (► Fig. 39.9).

39.10 Conclusion

Treatment of traumatic hand injuries in children can be a gratifying endeavor. Children heal well and exhibit better recovery from most injuries to the hand. Functional outcome is far superior to similar injuries in adults. However, the technical repair of these injuries can be challenging, as the structures are small.

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Part VI

Integument

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40 Vascular Anomalies

Arin K. Greene

Summary

Vascular anomalies are common lesions that affect the pediatric population. The most common tumors include infantile hemangioma, congenital hemangioma, kaposiform hemangioendothelioma, and pyogenic granuloma. The most frequent malformations are capillary malformation, venous malformation, lymphatic malformation, and arteriovenous malformation. Management is based on the type of lesion and may include laser, sclerotherapy, embolization, resection, or pharmacotherapy (topical, intralesional, systemic).

Keywords: vascular, anomaly, tumor, hemangioma, malformation

40.1 Introduction

Vascular anomalies are disorders that affect capillaries, arteries, veins, or lymphatics. Because these lesions usually involve the skin, the initial consultation often is with a plastic surgeon. Vascular anomalies are common, affecting 5.5% of the population. Lesions are classified biologically, based on their clinical behavior and cellular characteristics (► Table 40.1). The field is confusing because lesions look similar, and imprecise terminology is used (► Table 40.2).

There are two broad types of vascular anomalies: tumors and malformations. Tumors demonstrate endothelial proliferation and affect approximately 5% of the population. There are four major lesions: (1) infantile hemangioma (IH), (2) congenital hemangioma (CH), (3) kaposiform hemangioendothelioma

(KHE), and (4) pyogenic granuloma (PG; ► Fig. 40.1). Vascular malformations are errors in vascular development and have minimal endothelial turnover; they affect approximately 0.5% of the population. There are four major types based on the anomalous vessel(s): (1) capillary malformation (CM), (2) lymphatic malformation (LM), (3) venous malformation (VM), (4) arteriovenous malformation (AVM; ► Fig. 40.2). Recently, the causative mutation for many vascular anomalies has been identified (► Fig. 40.3).

40.2 Diagnosis

More than 90% of vascular anomalies can be diagnosed by history and physical examination (► Table 40.3). Approximately 9% of lesions require imaging for diagnosis, and less than 1% of vascular anomalies need histopathology to identify the lesion. Generally, the first-line imaging modality to confirm the type of vascular anomaly is ultrasound because it does not require sedation and is easy to perform. Magnetic resonance imaging (MRI) is used for lymphatic, venous, and AVMs to confirm their diagnosis and determine the extent of disease. Biopsy is reserved for a vascular anomaly that cannot be diagnosed despite imaging.

40.2.1 Infantile Hemangioma

IH is the most common tumor of infancy, affecting 4 to 5% of infants. IH is more frequent in premature children and in

Table 40.1 Classification of vascular anomalies

Tumors	Malformations		
	<i>Slow flow</i>	<i>Fast flow</i>	<i>Overgrowth syndromes</i>
Infantile hemangioma	Capillary malformation	Arteriovenous malformation	CLOVES
Congenital hemangioma	Venous malformation		Klippel-Trénaunay
Kaposiform hemangioendothelioma	Lymphatic malformation		Parkes Weber
Pyogenic granuloma			Sturge-Weber

Abbreviation: CLOVES, Congenital Lipomatosis Overgrowth, Vascular malformations, Epidermal nevi, and Scoliosis.

Table 40.2 Incorrect terminology commonly used to describe vascular anomalies

Tumors		Malformations	
<i>Biological name</i>	<i>Incorrect term</i>	<i>Biologic name</i>	<i>Incorrect term</i>
Infantile hemangioma	“Strawberry hemangioma” “Capillary hemangioma” “Cavernous hemangioma”	Capillary malformation	“Port-wine stain” “Capillary hemangioma”
Congenital hemangioma	“Infantile hemangioma”	Lymphatic malformation	“Cystic hygroma” “Lymphangioma”
Kaposiform hemangioendothelioma	“Capillary hemangioma”	Venous malformation	“Cavernous hemangioma”
Pyogenic granuloma	“Hemangioma”	Arteriovenous malformation	“Arteriovenous hemangioma”



Fig. 40.1 Examples of the four major types of vascular tumors. (a) Infantile hemangioma. (b) Congenital hemangioma. (c) Kaposiform hemangioendothelioma. (d) Pyogenic granuloma.



Fig. 40.2 Examples of the four major types of vascular malformations. (a) Capillary malformation. (b) Lymphatic malformation. (c) Venous malformation. (d) Arteriovenous malformation.



Fig. 40.3 Differentiating infantile hemangioma from a vascular malformation. (a-d) Infantile hemangioma grows rapidly during the first few months of life and then involutes. (e-h) Venous malformation of the lip. Vascular malformations are present at birth, slowly worsen, and do not regress.

Table 40.3 Mutations associated with vascular anomalies

Condition	Mutated gene	Inheritance
Capillary malformation	<i>GNAQ</i>	Somatic
Sporadic venous malformation	<i>TIE2</i>	Somatic
Verrucous venous malformation	<i>MAP3K3</i>	Somatic
Glomuvenous malformation	<i>Glomulin</i>	Dominant
Cutaneomucosal venous malformation	<i>TIE2</i>	Dominant
Cerebral cavernous malformation	<i>KRIT1</i>	Dominant
Sporadic lymphatic malformation	<i>PIK3CA</i>	Somatic
Familial congenital primary lymphedema	<i>VEGFR3</i>	Dominant
Lymphedema-distichiasis	<i>FOXC2</i>	Dominant
Lymphedema-hypotrichosis-telangiectasia	<i>SOX18</i>	Recessive
Hennekam syndrome	<i>CCBE1</i>	Recessive
Capillary malformation–arteriovenous malformation	<i>RASA1</i>	Dominant
Hereditary hemorrhagic telangiectasia type 1 (HHT1)	<i>ENG</i>	Dominant
Hereditary hemorrhagic telangiectasia type 2 (HHT2)	<i>ACVRLK1</i>	Dominant
PTEN-associated vascular anomaly	<i>PTEN</i>	Dominant

Abbreviation: PTEN, phosphatase and tensin homolog.

females (4:1). The median age of appearance is 2 weeks; 50% are noted at birth as a telangiectatic stain, pale spot, or ecchymotic area. IH grows faster than the child during the first 9 months of age (proliferating phase); 80% of its size is achieved by 3.2 (± 1.7) months. IH is red when it involves the superficial dermis. A lesion beneath the skin may not be appreciated until 3 to 4 months of age when it has grown large enough to cause a visible mass; the overlying skin may appear bluish. After 12 months, the tumor begins to regress (involuting phase); the color fades and the lesion flattens. Involution ceases in most of children by age 4 years (involved phase). After involution, one-half of children will have residual telangiectasias, scarring, fibrofatty residuum, redundant skin, or destroyed anatomical structures.

Infants with five or more small (<5 mm) IHS have a 16% risk of having hepatic hemangiomas, which are typically asymptomatic. PHACE association consists of a plaque-like IH in a regional distribution of the face with at least one of the following anomalies: Posterior fossa brain malformation, Hemangioma, Arterial cerebrovascular anomalies, Coarctation of the aorta and cardiac defects, Eye/Endocrine abnormalities. Because 8% of children have a stroke in infancy, these patients should have an MRI to evaluate the brain and cerebrovasculature. Infants are referred for ophthalmologic, endocrine, and cardiac evaluation to rule out associated anomalies. LUMBAR association (Lower body infantile hemangioma, Urogenital anomalies, Myelopathy, Bony deformities, Anorectal malformations, Renal anomalies) typically affects the sacral area or lumbar region. Ultrasonography is obtained to rule out associated anomalies in infants less than 4 months of age. MRI is indicated in older infants or when ultrasound is equivocal.

40.2.2 Congenital Hemangioma

CHs are fully grown at birth and do not have postnatal growth. They are red-purple with coarse telangiectasias, central pallor, and a peripheral pale halo. These lesions are more common in

the extremities, have an equal sex distribution, and are solitary with an average diameter of 5 cm. There are two forms: rapidly involuting congenital hemangioma (RICH) and noninvoluting congenital hemangioma (NICH). RICH involutes rapidly after birth and 50% complete regression by 7 months of age; the remaining tumors are fully involuted by 14 months. NICH, in contrast, does not regress and remains unchanged.

40.2.3 Kaposiform Hemangioendothelioma

KHE is a rare neoplasm (1/100,000 children) that does not metastasize. KHE is present at birth in 50% of patients, has an equal sex distribution, and affects the head/neck (40%), trunk (30%), or an extremity (30%). The tumor is often greater than 5 cm and is reddish-purple. Seventy percent of patients have Kasabach–Merritt phenomenon (KMP; thrombocytopenia < 25,000/mm³, petechiae, bleeding). KHE partially regresses after 2 years of age, although it usually causes chronic pain and stiffness. MRI is indicated for diagnostic confirmation and to assess the extent of the tumor.

40.2.4 Pyogenic Granuloma

PG is a solitary, red papule that grows rapidly on a stalk. It is small, with an average diameter of 6 mm; the mean age of onset is 6 years. PG commonly is complicated by bleeding (64%) and ulceration (36%). PG involves the skin (88%) or mucous membranes (11%). It is distributed on the head or neck (62%), trunk (19%), upper extremity (13%), or lower extremity (5%). In the head and neck region, affected sites include cheek (29%), oral cavity (14%), scalp (11%), forehead (10%), eyelid (9%), or lips (9%).

40.2.5 Capillary Malformation

CM (previously called “port-wine stain”) is the most common type of vascular malformation. The lesion is noticed at birth and can involve any area of the integument. Over time, the lesion progresses: (1) it darkens, (2) fibrovascular cobblestoning can occur, (3) PGs may develop, and (4) soft-tissue and bony may enlarge underneath the stain. The birthmark referred to as an “angel kiss” or “stork bite” is a fading capillary stain. It is present in one-half of Caucasian newborns and is located on the forehead, eyelids, nose, upper lip, or posterior neck. No treatment is necessary because it lightens over the first 2 years of life.

40.2.6 Venous Malformation

Although VMs are present at birth, they may not become evident until childhood or adolescence when they have grown large enough to cause a visible deformity or symptoms. Lesions are blue, soft, and compressible. Hard, calcified phleboliths may be palpable. The primary morbidity is psychosocial because most lesions affect the skin and cause a deformity. The second most common complication is pain secondary to thrombosis and phlebolith formation. Patients are not at risk for thromboembolism, unless a large phlebectatic vein is connected to the deep venous system. VMs are diagnosed by history and

physical examination. Dependent positioning will cause a lesion to enlarge. Small, superficial VMs do not require further diagnostic workup. Large or deep lesions are evaluated by MRI. Approximately 10% of patients with VM have multifocal, familial lesions that are autosomal dominant: glomuvenous malformation (GVM), cutaneomucosal-venous malformation (CMVM), or cerebral cavernous malformation (CCM).

Several phenotypical subtypes of VM exist. Blue rubber bleb nevus syndrome (BRBNS) is characterized by multiple, small VMs involving the skin, soft tissue, and gastrointestinal tract. Morbidity is associated with gastrointestinal bleeding. Diffuse phlebectasia of Bockenhimer specifies an extensive extremity VM involving skin, subcutaneous tissue, muscle, and bone. Sinus pericranii refers to a venous anomaly of the scalp or face and transcalvarial communication with the dural sinus. Verrucous VM is clinically similar to a hyperkeratotic VM. Fibroadipose vascular anomaly is differentiated from intramuscular VM by significant pain, contractures, and a solid component.

40.2.7 Lymphatic Malformation

LM is defined by the size of its channels: macrocystic, microcystic, or combined. Lesions are usually noted at birth, although small or deep LMs may not become evident until childhood or adolescence after they have enlarged and/or become symptomatic. The most commonly affected sites are the neck and axilla. LM causes three major problems: (1) psychosocial morbidity, (2) infection, and (3) bleeding. Macrocystic lesions contain cysts large enough to be accessed by a needle (typically ≥ 5 mm) and are amenable to sclerotherapy. The most commonly affected sites are the neck or axilla. Microcystic LMs have cysts that are too small to be cannulated by a needle and thus cannot be treated by sclerotherapy. These lesions commonly affect the face and extremities and are often associated with cutaneous vesicles that can bleed and leak lymph fluid. Approximately one-half of LMs are not purely macrocystic or microcystic; they contain both macrocysts and microcysts. Primary lymphedema (affecting 1/10,000 persons) is a type of LM. Ninety percent of LMs are diagnosed by history and physical examination. Small, superficial LMs do not require further diagnostic workup. Large or deep lesions are evaluated by MRI.

40.2.8 Arteriovenous Malformation

AVM has an absent capillary bed, which causes shunting of blood directly from the arterial to venous circulation through a fistula (direct connection of an artery to a vein) or nidus (abnormal channels bridging the feeding artery to the draining veins). Although present at birth, an AVM may not become evident until childhood, after it has enlarged or become symptomatic. Lesions have a pink-red cutaneous stain, are warm, and can have palpable pulsations. Patients are at risk for pain, ulceration, bleeding, and congestive heart failure. AVMs also can cause disfigurement, destruction of tissues, and obstruction of vital structures. Ninety percent of AVMs are diagnosed by history and physical examination. Hand-held Doppler examination shows fast flow. If the diagnosis is equivocal after history and physical examination, ultrasound is the first-line study to confirm the diagnosis. MRI usually is obtained to determine the extent of the lesion.

Capillary malformation-arteriovenous malformation (CM-AVM) is an autosomal-dominant condition. Patients have atypical CMs that are small, multifocal, round, pinkish-red, and often surrounded by a pale halo (50%). One-third of patients also have an AVM: Parkes Weber syndrome (PWS; 12%), extracerebral AVM (11%), or intracerebral AVM (7%). A patient presenting with multiple CMs, especially with a family history of similar lesions, should be evaluated for a possible AVM. Because 7% of patients with CM-AVM will have an intracranial fast-flow lesion, brain MRI should be considered. Exploratory imaging of other anatomical areas is not necessary because extracranial AVMs have not been found to involve the viscera.

40.3 Nonoperative Management

40.3.1 Infantile Hemangioma

Most IHs are observed because they are small, localized, and do not involve anatomically important areas. During the proliferative phase, 16% ulcerate at a median age of 4 months; the lips, neck, and anogenital region are the most common locations. To protect against ulceration, IH in these areas should be kept moist with hydrated petroleum during the proliferative phase to minimize desiccation and shearing of the skin. If an ulceration develops, the wound is washed gently with soap and water at least twice daily. Small, superficial areas are managed by the application of topical antibiotic ointment and occasionally with a petroleum gauze barrier. Large, deep ulcers require

damp-to-dry dressing changes. To minimize discomfort, a small amount of topical lidocaine may be applied no more than four times daily to avoid toxicity. Bleeding from an ulcerated IH is usually minor, and is treated by applying direct pressure. Ulcerations will heal with local wound care within 2 to 3 weeks.

40.3.2 Topical Pharmacotherapy

Topical corticosteroid is relatively ineffective, especially if IH involves the deep dermis and subcutis. Although lightening may occur, if there is deep component it will not be affected. Adverse effects include hypopigmentation and cutaneous atrophy. Topical timolol may be effective for superficial lesions, but will not affect IHs with a subcutaneous component.

40.3.3 Intralesional Corticosteroid

Problematic IHs that are well localized and less than 3 cm are best managed by intralesional corticosteroid (► Fig. 40.4). Triamcinolone (not to exceed 3 mg/kg) will stop the growth of the lesion; two-thirds will decrease in size. The corticosteroid lasts 2 to 3 weeks, and thus infants may require two to three injections during the proliferative phase.

40.3.4 Systemic Pharmacotherapy

Problematic IHs greater than 3 cm are managed by oral prednisolone or propranolol (► Fig. 40.5). Prednisolone has been used

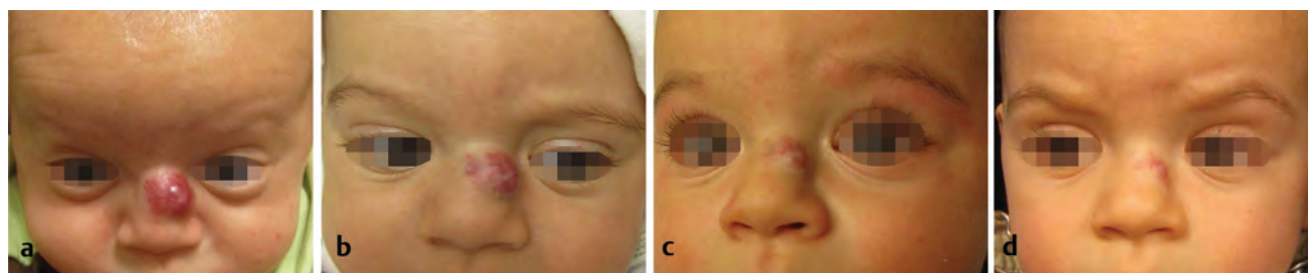


Fig. 40.4 Management of a localized proliferating infantile hemangioma with intralesional corticosteroid. (a) Three months of age at time of kenalog injection. (b) Accelerated regression of the tumor at 4 months of age. (c) Age 8 months. (d) Age 12 months.



Fig. 40.5 Treatment of a large proliferating infantile hemangioma with oral prednisolone. Note accelerated regression of the tumor.

to treat IH for over 40 years and has proven to be very safe and effective. Patients are given 3 mg/kg/d for 1 month; the drug is then tapered by 0.5 mL every 2 to 4 weeks until it is discontinued between 10 and 12 months of age when the tumor is no longer proliferating. The drug is given once a day in the morning, and infants have monthly outpatient follow-up. Using this protocol, all tumors will stabilize in growth and 88% will become smaller (accelerated regression). Twenty percent of infants will develop a cushingoid appearance that resolves during tapering of therapy. Approximately 12% exhibit decreased gain in height, but return to their pretreatment growth curve by 24 months of age.

Propranolol dosing typically is 2 mg/kg/d. Approximately 90% of tumors will stop growing or regress. Risks (<3%) include bronchospasm, bradycardia, hypotension, hypoglycemia, seizures, and hyperkalemia. Preterm infants and those less than 3 months of age are more likely to have adverse events. Patients usually have cardiology consultation, electrocardiogram, echocardiogram, glucose/electrolyte measurements, and frequent blood pressure, heart rate, and respiratory examinations. Inpatient initiation of treatment is used for premature or infants less than 3 months of age. Potential contraindications include asthma, glucose abnormalities, heart disease, hypotension, bradycardia, PHACES association. The drug should be discontinued if the infant is ill because reduced oral intake can increase the risk of hypoglycemia and seizures. Patients treated with propranolol experience later rebound growth compared to prednisolone, and thus may necessitate longer treatment. Recently, concern has been raised about potentially negative long-term neurocognitive effects of propranolol when given to infants.

40.3.5 Laser Therapy

Pulsed dye laser treatment generally is not indicated for proliferating IH. The laser penetrates only approximately 1 mm into the dermis, and thus only affects the superficial portion of the tumor. Although lightening may occur, the mass is not affected. Patients have an increased risk of skin atrophy, ulceration, pain, bleeding, scarring, and hypopigmentation. Pulsed dye laser is indicated during the involuted phase to fade residual telangiectasias.

40.3.6 Congenital Hemangioma

RICH usually does not require intervention during infancy because it regresses quickly. Rarely, it can cause congestive heart failure, which is treated with corticosteroid or embolization. NICH is rarely problematic in infancy and is observed.

40.3.7 Kaposiform Hemangioendothelioma

Most lesions are extensive and are unable to be resected. Vincristine is first-line therapy; the response rate is 90%. Recently, patients have been treated with sirolimus with good efficacy. Thrombocytopenia is not significantly improved with platelet transfusion, which should be avoided unless there is active bleeding or a surgical procedure is planned. By 2 years of age,

the tumor has undergone partial involution and the platelet count normalizes.

40.3.8 Pyogenic Granuloma

Because PG frequently bleeds, lesions are covered to avoid incidental trauma. Bleeding is controlled by application of pressure. Rarely, a PG can “fall off” and not recur. However, almost all lesions require resection.

40.3.9 Capillary Malformation

Patients with CMs in nonvisible locations may not elect for intervention. The mainstay of treatment is pulsed dye laser, which lightens its color. Intervention during infancy or early childhood is recommended because (1) superior lightening of the lesion is achieved, (2) the risk of darkening and hypertrophy is reduced, and (3) psychosocial morbidity is minimized. Pulsed dye laser is less effective for CMs that have progressed to a dark color with cutaneous thickening.

40.3.10 Venous Malformation

VM can be observed; intervention is reserved for symptomatic lesions or asymptomatic areas at risk for thromboembolism. Patients with an extensive extremity VM are prescribed custom-fitted compression garments. Individuals with recurrent pain secondary to phlebothrombosis are given aspirin to prevent thrombosis. Disseminated intravascular coagulopathy (DIC) can occur following trauma or interventions. Chronic consumptive coagulopathy can cause thrombosis (phleboliths) or bleeding (hemarthrosis, hematoma, intraoperative blood loss). Low molecular weight heparin is considered for patients at risk for DIC. Individuals who develop a serious thrombotic event require long-term anticoagulation or a vena caval filter.

40.3.11 Lymphatic Malformation

LM is benign and thus intervention is not mandatory. Intraleisional bleeding is treated with pain medication. Patients with more than three infections a year are given prophylactic antibiotics. First-line management for problematic macrocystic/combined LM is sclerotherapy. This technique involves aspiration of the cysts followed by the injection of an inflammatory substance, which causes scarring of the cyst walls to each other. Sclerotherapy shrinks the lesion and alleviates symptoms; it gives superior results and has lower morbidity compared to resection. Oral sirolimus is now being used to treat problematic microcystic LMs that have failed or are not amenable to other interventions.

40.3.12 Arteriovenous Malformation

AVM does not necessarily require intervention. Because the lesion is often diffuse and involves multiple tissue planes, cure is rare. An asymptomatic AVM should be observed unless it can be removed for possible cure with minimal morbidity.

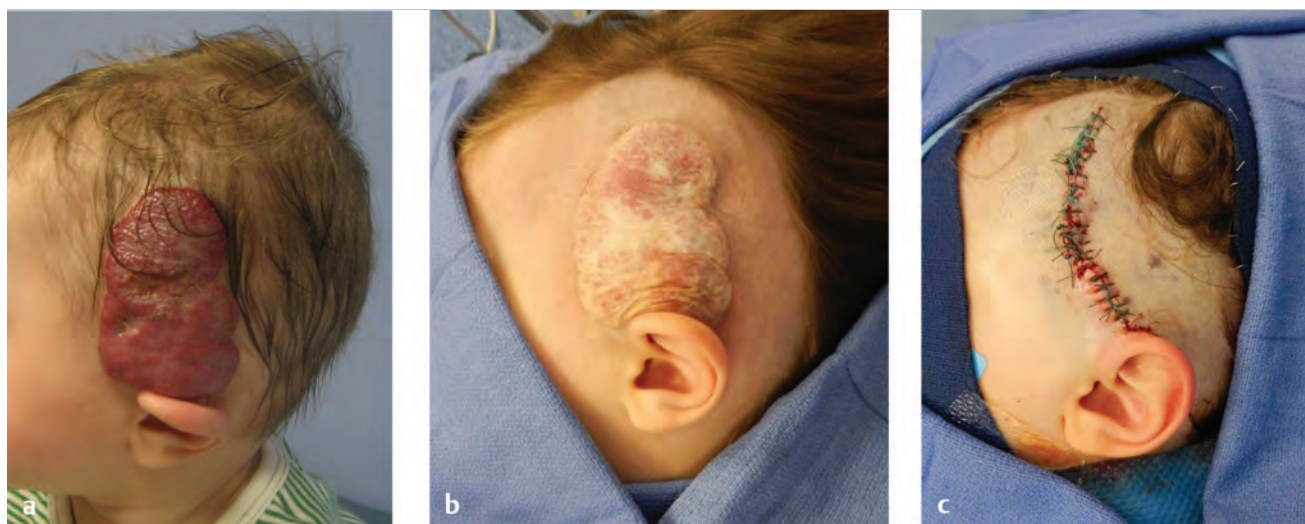


Fig. 40.6 Resection of infantile hemangioma. It is preferable to wait until the tumor has regressed (> 3 years of age) before operative intervention. (a) Age 1 year. (b) Age 3 years at time of resection. (c) Following removal of the involuted lesion.

40.4 Operative Treatment

40.4.1 Infantile Hemangioma

Resection generally is not recommended during the proliferating phase. The tumor is highly vascular, and there is a risk for blood loss, iatrogenic injury, and an inferior outcome, compared to excising residual tissue after the tumor has regressed. Approximately 50% of IH leave behind fibrofatty tissue or damaged skin after the tumor regresses, causing a deformity. It is preferable to intervene surgically between 3 and 4 years of age (► Fig. 40.6). During this period, the IH will no longer improve significantly, and the procedure is performed before the child's long-term memory and self-esteem begin to form at about 4 years of age. Also, waiting until the IH is at its smallest will result in the shortest possible scar and the most favorable outcome. Circular lesions located in visible areas, particularly the face, are best removed by circular excision and purse-string closure. This technique minimizes the length of the scar as well as distortion of surrounding structures. Lenticular excision of a circular IH results in a scar as long as three times the diameter of the lesion. In comparison, a two-stage circular resection followed by lenticular excision/linear closure will leave a scar approximately the same length as the diameter of the original lesion.

40.5 Congenital Hemangioma

After regression, RICH can leave behind atrophic skin and subcutaneous tissue. Reconstruction with autologous grafts (fat, dermis) or acellular dermis may be indicated. Resection of NICH may be indicated to improve the appearance of the affected area, as long as the surgical scar will be less noticeable than the lesion.

40.5.1 Kaposiform Hemangioendothelioma

KHE typically involves multiple tissue planes and is too diffuse to resect. Rarely, if a lesion is localized and amenable to

operative intervention, excision should be considered to obviate the need for systemic chemotherapy.

40.5.2 Pyogenic Granuloma

PGs require intervention to control likely ulceration and bleeding. Because PG extends into the reticular dermis, the lesion usually is out of the reach of the pulsed dye laser, cautery, or shave excision. Consequently, these modalities have a recurrence rate of approximately 50%. Full-thickness excision is definitive treatment. Superficial, small lesions can be treated with needle-tip cautery through the reticular dermis.

40.5.3 Capillary Malformation

CMs typically do not require operative intervention. Surgical procedures are indicated to correct overgrowth caused by the malformation (► Fig. 40.7). Small fibrovascular nodules or PGs can be excised. Trunk or extremity CM associated with increased subcutaneous adipose tissue can be improved with suction-assisted lipectomy. Severe cutaneous thickening and cobblestoning may be resected and reconstructed by linear closure, skin grafts, or local flaps. Malocclusion can be corrected in adolescence with orthodontics; an orthognathic procedure is considered when the jaws are completely grown. Facial asymmetry caused by overgrowth of the zygoma, maxilla, or mandible can be improved by contour burring.

40.5.4 Venous Malformation

First-line treatment is sclerotherapy, which is safer and more effective than resection (► Fig. 40.8). Diffuse lesions are managed by targeting specific symptomatic areas. Sclerotherapy is repeated until symptoms are alleviated or when injectable vascular spaces are no longer present. Although sclerotherapy effectively reduces the size of the lesion and improves symptoms, the malformation remains. Consequently, patients may have a mass or visible deformity after treatment that may be

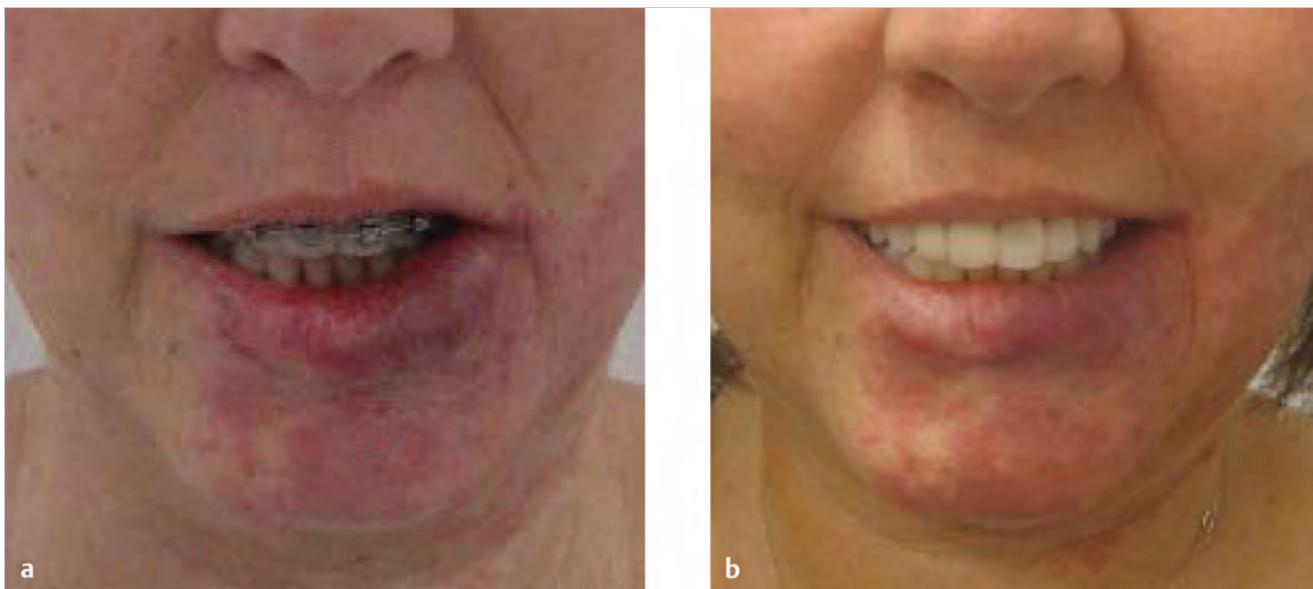


Fig. 40.7 Management of a capillary malformation with pulsed dye laser and transverse mucosal resection of the left lower lip. **(a)** Prior to intervention. **(b)** Postoperative appearance.



Fig. 40.8 Intervention for venous malformation. **(a-c)** Adolescent female with new onset swelling of the left mandible managed with sclerotherapy. **(d,e)** Resection of a localized venous malformation of the scalp.

improved by resection. VM usually re-expands after sclerotherapy, and thus patients often require additional treatments. Our preferred sclerosant is sodium tetradecyl sulfate. Most patients, especially children, are managed under general anesthesia using imaging.

In contrast to sclerotherapy, resection is rarely primary treatment because (1) the entire lesion is difficult to remove; (2) the risk of recurrence is high because hidden channels adjacent to the visible lesion are not excised; (3) the risk of blood loss and iatrogenic injury is greater. Resection should be considered for (1) small, well-localized lesions that can be completely removed or (2) persistent mass or deformity after completion of sclerotherapy (patent channels are no longer accessible for further injection). When considering resection, the postoperative scar/deformity should be weighed against the preoperative appearance of the lesion. Subtotal resection of a problematic area is indicated rather than attempting “complete” excision of a benign lesion that might result in a worse deformity than the malformation itself. Almost all VMs should have sclerotherapy prior to operative intervention. After adequate sclerotherapy,

the VM is replaced by scar and thus the risk of blood loss, iatrogenic injury, and recurrence is reduced. In addition, fibrosis facilitates resection and reconstruction.

40.5.5 Lymphatic Malformation

Sclerotherapy is first-line management for large or problematic macrocystic/combined LM (we prefer doxycycline; ► Fig. 40.9). Recently, bleomycin sclerotherapy has shown efficacy for microcystic LMs. This modality can be considered for problematic lesions where resection would be unfavorable; a 10 to 25% reduction in the size of the lesion can be expected. Cutaneous LMs that are bleeding or leaking lymph fluid can be treated with carbon dioxide laser. Radiofrequency ablation can improve intraoral lymphatic vesicles.

Resection is reserved for (1) symptomatic microcystic LM causing bleeding, infection, distortion of vital structures, or significant deformity; (2) symptomatic macrocystic/combined LM that no longer can be managed with sclerotherapy because all macrocysts have been treated; (3) small, well-localized LM



Fig. 40.9 Intervention for lymphatic malformation. (a,b) Child with a macrocystic lesion of the left upper eyelid treated with sclerotherapy. (c,d) Resection of a microcystic lymphatic malformation of the lower extremity.

(microcystic or macrocystic) that may be completely excised. When considering resection, the postoperative scar/deformity should be weighed against the preoperative appearance of the lesion. For diffuse LMs, staged resection of defined anatomic areas is recommended. Subtotal excision of problematic areas, such as bleeding vesicles or a hypertrophied lip, should be carried out rather than attempting to “complete” resection that might result in a worse deformity than the malformation itself. Bleeding or leaking cutaneous vesicles can be controlled by resection if they are localized. Large areas of vesicular bleeding or drainage are best managed by sclerotherapy or carbon dioxide laser; alternatively, wide resection and skin graft coverage is required. Microcystic vesicles involving the oral cavity respond well to radiofrequency ablation.

40.5.6 Arteriovenous Malformation

Embolization is generally the first-line treatment. The procedure involves the delivery of a substance through a catheter into the AVM nidus to occlude blood flow and/or fill a vascular space. Scarring reduces arteriovenous shunting, shrinks the lesion, and diminishes symptoms. Embolization is used either as a preoperative adjunct to resection or as monotherapy for lesions not amenable to extirpation. Despite the high likelihood of re-expansion, embolization can effectively palliate an AVM by reducing its size, slowing enlargement, and alleviating pain and bleeding. The embolic material is delivered to the nidus, not to the proximal arterial feeding vessels. Occlusion of inflow will cause collateralization and expansion of the AVM; access to the nidus also will be blocked, preventing future embolization.

Resection of AVM has a lower recurrence rate than embolization alone; it is considered for a well-localized lesion or to correct deformity (i.e., bleeding or ulcerated areas, labial hypertrophy; ► Fig. 40.10). Wide extirpation and reconstruction of

large, diffuse AVM should be exercised with caution because (1) cure is rare and the recurrence rate is high; (2) the resulting deformity is often worse than the appearance of the malformation; (3) resection is associated with significant blood loss, iatrogenic injury, and morbidity. When excision is planned, preoperative embolization will facilitate the procedure by reducing the size of the AVM, minimizing blood loss, and creating scar tissue to aid the dissection. Excision should be done 24 to 72 hours after embolization, before recanalization restores blood flow to the lesion.

40.6 Vascular Malformation Overgrowth Syndromes

CLOVES Syndrome (Congenital Lipomatosis Overgrowth, Vascular malformations, Epidermal nevi, and Scoliosis)

All patients have a truncal lipomatous mass, a slow-flow vascular malformation (most commonly a CM overlying the lipomatous mass), and hand/foot anomalies (increased width, macrodactyly, first webspace sandal gap). Patients also may have AVM (28%), neurological impairment (50%), or scoliosis (33%).

40.6.1 Klippel–Trénaunay Syndrome

This is a capillary-lymphatic-VM of an extremity causing overgrowth. The condition almost always affects the lower limb. A large, embryonic vein in the subcutaneous tissue isolated in the lateral calf/thigh and communicates with the deep venous system. Complications include thrombophlebitis and pulmonary embolism. The lymphatic abnormalities are usually macrocystic in the pelvis/thigh and microcystic in the abdominal wall, buttock, and distal limb. Because embryonal veins can connect to



Fig. 40.10 Resection of an arteriovenous malformation. Child with a bleeding, ulcerated lesion of the scalp managed with preoperative embolization followed by excision.

Table 40.4 Management summary for the major types of vascular anomalies

Tumors		Malformations	
<i>Biological name</i>	<i>Treatment</i>	<i>Biological name</i>	<i>Treatment</i>
Infantile hemangioma	Observe Topical timolol Kenalog injection Oral prednisolone Oral propranolol Resection	Capillary malformation	Observe Laser Resection
Congenital hemangioma	Observe Resection	Lymphatic malformation	Observe Sclerotherapy Resection
Kaposiform hemangioendothelioma	Vincristine Sirolimus	Venous malformation	Observe Sclerotherapy Resection
Pyogenic granuloma	Resection	Arteriovenous malformation	Observe Embolization Resection

the deep venous system causing thromboembolism, they are removed in early childhood with sclerotherapy or endovascular laser.

40.6.2 Parkes Weber Syndrome

PWS consists of a diffuse AVM of an extremity (usually the leg) causing soft-tissue and/or bony overgrowth. A CM involves the skin of the affected limb. Patients have subcutaneous and intramuscular microshunting and can develop congestive heart failure. Most children are observed until symptoms necessitate intervention. Embolization may reduce congestive heart failure, pain, or ulceration. Occasionally, amputation is necessary.

40.6.3 Phosphatase and Tensin Homolog Hamartoma-Tumor Syndrome

Patients with phosphatase and tensin homolog (*PTEN*) mutations, a tumor suppressor gene, have PTEN hamartoma-tumor syndrome (PHTS), an autosomal dominant condition. Approximately 50% of patients have a unique fast-flow vascular anomaly with arteriovenous shunting, referred to as a PTEN-associated vascular anomaly. PHTS also is associated with autism (19%), thyroid lesions (31%), or gastrointestinal polyps (30%).

40.6.4 Sturge–Weber Syndrome

The Sturge–Weber syndrome (SWS) is defined by (1) a CM in the V₁ trigeminal nerve distribution with either (2) ocular abnormalities (glaucoma, choroidal vascular anomalies) and/or (3) a leptomeningeal vascular malformation. Pial vascular lesions may cause refractory seizures, hemiplegia, and /or delayed cognitive development. Any child with a CM in an upper trigeminal nerve distribution should be screened for SWS. MRI is obtained to rule out leptomeningeal vascular lesions. Patients undergo ophthalmology evaluation to assess for choroidal anomalies and glaucoma.

40.7 Conclusion

The field of vascular anomalies is confusing because lesions look similar and terminology is difficult. More than 90% of patients can be diagnosed by history and physical examination. Treatment of a vascular anomaly is not considered until the diagnosis is confirmed. An experienced physician can manage most vascular anomalies independently. However, many lesions require interdisciplinary care and should be referred to a vascular anomalies center. Although vascular tumors are at least 10 times more common than vascular malformations, vascular malformations are generally more problematic and comprise two-thirds of patients managed in a vascular anomalies center. If the clinician initially focuses on the major types of vascular anomalies, then he/she will be able to manage approximately 95% of patients correctly (► Table 40.4).

40.8 Key Points

- Vascular anomalies are divided into tumors or malformations.
- IH is the most common tumor of infancy; it grows rapidly after birth and involutes during childhood.
- Most IHS are observed; problematic lesions are treated pharmacologically or by resection.
- Vascular malformations are present at birth, although not always obvious; they may slowly enlarge during childhood and adolescence.
- Vascular malformations are managed by observation, laser, sclerotherapy, embolization, or resection; pharmacotherapy is not available.

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41 Burns

Matthias B. Donelan and Branko Bojovic

Summary

Reconstruction of pediatric burns have improved recently. Management is based on the principles of wound healing and scar maturation. Tissue can be manipulated with topical and intralesional modalities, local tissue rearrangements, grafts, and photomedicine.

Keywords: pediatric, burn, scar, reconstruction, skin graft, laser

41.1 Introduction

There has never been a more exciting and optimistic time in the history of pediatric burn therapy than that exists today in the developed world. Current therapeutic options to treat children who sustain burn injuries of all varieties have now reached a state of effectiveness that was unimaginable only a few short decades ago. Progress in the understanding of wound healing, improved diagnosis through technological advances, vastly superior critical care, as well as new and refined techniques of reconstructive surgery make the care of pediatric burns and restoration of the patients' quality of life an exciting and rewarding field of pediatric plastic surgery. Because burns can involve all areas of the body and vary in size from the very small to almost the entire body surface area, they require a completely competent and caring plastic surgeon to treat the whole patient, the whole family, and the whole range of deformities that can be encountered. A few salient strategic concepts, however, are worth emphasizing at the beginning of this chapter.

Acute burn injuries that are not life-threatening should be cared for with the most conservative burn wound management possible. Early excision and grafting commits a patient to a donor site and a grafted outcome that is iatrogenic and permanent (► Fig. 41.1). When necessary, to prevent serious complications, such early surgical intervention is essential. When not necessary, it is harmful for the patient. Improved diagnostic techniques such as laser Doppler imaging have been validated in multiple large studies and should be incorporated in the care of all pediatric burn injuries if at all possible. The overwhelming consensus from recent literature reports originated in the United Kingdom and Belgium has demonstrated that following the adaptation of laser Doppler imaging, the incidence of early excision and grafting in pediatric plastic surgery units has significantly decreased. Early excision and grafting has been advocated in smaller burns because of an alleged superior outcome and prevention of hypertrophic scarring. Profound advances in our ability to prevent and treat hypertrophic scarring as well as combining local plastic surgery techniques and the use of lasers and other photomedicine have allowed us to regenerate scars in ways that were inconceivable as recently as the 1990s. Today, a patient's hypertrophic, contracted scars are often their most valuable reconstructive donor sites. The reason for that is that nothing beats "original equipment," and their own scarred

tissue is potential reconstructive material in the right location with the best possible color and texture match. It also does not require the creation of an iatrogenic donor site deformity on an uninjured part of the patient's body (► Fig. 41.2). The ability to help scars to regenerate themselves is an essential part of pediatric burn care and reconstruction.

41.2 Diagnosis

Burn deformities present unique and complex reconstructive challenges for the pediatric plastic and reconstructive surgeon. A thorough understanding of the burn wound and its evolution is critical to successful treatment outcomes and is part of sound surgical judgment and expertise. Burn care can be considered as one of the most challenging areas of medicine because it involves multiple medical and surgical treatment modalities



Fig. 41.1 A 4-year-old female who sustained a small, mixed second-degree scald burn at age 18 months. Early excision and meshed skin grafts resulted in a permanent, severe cosmetic deformity.



Fig. 41.2 (a) A 4-year-old female from Mongolia who sustained second- and third-degree flame burns at age 2 in yurt (ger) fire. Wounds healed by contraction and epithelialization, resulting in severe contractures and hypertrophic scarring. (b) Contractures were relieved by Z-plasties using the contracted, hypertrophic scar tissue. (c) Three years after three Z-plasty procedures and six treatments with the fractionated CO₂ laser and topical triamcinolone (10 mg/mL). The contractures are completely released and normal contour is restored.



Fig. 41.3 (a) An 11-year-old female 18 months following a flash burn to the face resulting in diffuse hypertrophic scarring, contractures, and ulceration, as well as severe pruritus. (b) Contractures were relieved and contours leveled with multiple Z-plasties during four separate procedures over 6 years. Scars were also treated simultaneously nine times with the pulsed dye laser and six times with the fractionated, ablative CO₂ laser. No scars were excised. (c) Six years after initial treatment, her face is normal except for slight textural irregularity and mild dyspigmentation.

implemented over the course of time, frequently many years. It is beyond the scope of this chapter to discuss acute burn care, but suffice to say that well-described methods for recognizing burn injury from a number of modalities are replete within the literature. Thermal, electrical, and chemical injuries form the basis of the majority of the causative factors for acute burn injuries.

Burns, in general, constrict and deform and in doing so alter contour, proportions, features, form, and function. Burn care

should strive to restore the injured patient as much as possible to their preburn condition and as close to “normal” as is possible with minimal morbidity (► Fig. 41.3). A return to normal, that is, the way in which the patient existed prior to the burn, is not possible. This must be clearly communicated to the patient and his or her loved ones so that they understand that the journey of burn care and reconstruction is one that sets realistic expectations and goals in treatment and offers the hope that we can get such patients to “near-normal.” Such an approach is

realistic, responsible, and still offers optimism and hope to the patient, and family, that he or she can look forward to a return as close to the preburn injury condition as is possible, save for what the future may hold with regenerative medicine or technological advancements on the horizon.

The ultimate goal of burn reconstruction should be the restoration of an aesthetically pleasing and absolutely tension-free appearance to allow for appropriate form and function. The keys to achieving that goal are to appropriately diagnose the burn scar deformity, understand what issues are preventing that goal, and then provide a sound plan to address the problems. In concept, this sounds simple, but in practice it requires expertise and resources to make this goal a reality. Controversy still exists on what the optimum treatment is for moderate to severe burns. The vast majority of burns treated conservatively with topical antibiotics should heal in approximately 3 weeks. Burns that are obviously full thickness are best treated by early, tangential excision and grafting within 7 to 10 days. Even for controversial areas, the majority of burns can and are treated expectantly in most pediatric burn centers with a combination of modalities, which will be addressed later in this chapter.

Several useful generalizations regarding diagnosis and correct characterization of burns are important to mention and keep in mind. Superficial second-degree burns typically heal without scarring or pigmentary changes to the skin. Deeper or medium-thickness second-degree burns that epithelialize in 10 to 14 days will often heal without scarring although long-term changes in skin pigmentation and texture are frequent occurrences. Deep second-degree burns that epithelialize in 14 to 28 days or longer require careful observation; they are prone to develop severe late hypertrophic scarring. These patients, in particular, require close and continuous follow-up after the initial healing phase and must be managed via all available ancillary modalities, including appropriately timed surgical intervention when indicated.

41.3 Nonoperative Management

Whether we are considering nonoperative or operative treatment of burns, both modalities are based on an overall strategy and understanding of the specific problem at hand. This was briefly mentioned already in the “Diagnosis” section. Many techniques, including a plethora of surgical ones, have been described in the literature to address burns of many varieties and depths. What is most important is that in all cases, fundamental principles are recognized and followed such that the goals of management and treatment are arrived at in a predictable fashion. Therefore, developing a strategic concept of burn treatment is paramount and will help avoid unfortunate results.

Deep second- and third-degree burns heal by contraction and epithelialization and the deeper the injury, the more healing takes place by contraction. This category of burn depth will be expanded upon in the operative treatment section to follow as these typically do not do well without operative management. More superficial burns, as mentioned previously, require patience, observation, and providing the wounded area a milieu

to allow for rapid, infection-free re-epithelialization. This is typically accomplished by good hygiene of the area, judicious pain control, and local wound care with a combination of modalities that include antibiotic topical creams or gels to more sophisticated wound dressings that are applied and changed daily or every several days while awaiting wound re-epithelialization. Typically, these burns wounds are those which heal within days to less than 2 weeks. Once the wound is re-epithelialized and risk of infection has passed, other well-described modalities exist for adjunctive therapy. Pressure garments have long been demonstrated to be effective in suppressing and reversing hypertrophic scarring. The addition of silicone to pressure therapy has also been a significant advance. Judicious use of sunscreen on immature and newly re-epithelialized burn scars is mandatory and typically should last a year as the scar undergoes maturation and is prone to hyperpigmentation from ultraviolet (UV) radiation. Sunscreen, in general, should be encouraged and recommended as part of a daily program of skin protection and treatment, regardless of burn depth and as part of normal pediatric skin care and skin maintenance.

41.4 Operative Treatment

As discussed earlier, a clear understanding of the pathophysiology responsible for a chronic open burn wound or burn scar contracture is essential to be able to devise an operative strategy to treat it. Several fundamental concepts and techniques are important to discuss prior to describing specific operative interventions: contractures, contour and proportion, tension, donor site availability, aesthetic units, Z-plasty, grafts, flaps, and lasers. These nine basic treatment strategies must be understood in order to have a comprehensive overview of operative pediatric burn wound management and treatment.

41.4.1 Contractures

Burn injury leads to tissue loss and the resultant wounds heal with contraction, which results in contractures. Contractures are either “intrinsic” or “extrinsic.” Intrinsic contractures are those that result from loss of tissue in the area of the contracture with subsequent distortion of the part affected. Extrinsic contractures include those in which the tissue loss is at a distance from the affected area, but the distorted structures are themselves not injured. It is critical to recognize these differences given that they are often overlooked in burn scar and burn injury treatment. Not only must corrective measures be directed at the cause of the contracture to prevent iatrogenic deformities, but it is also essential to minimize the amount of skin and scar that are excised when dealing with contractures. Minimizing excision decreases the amount of new skin that must be provided. In fact, due to the beneficial effects of releasing tension, most scars will mature favorably once a release has been carried out (► Fig. 41.2 and ► Fig. 41.3). It is important to understand that scars are living things, not static entities, and that they will respond to a change in their environment.



Fig. 41.4 (a) Severe, type II, pan-facial burn deformity in this 5-year-old female with significant loss of facial anatomy after a 70% total body surface area flame burn in a house fire. (b) After restoration of normal facial proportions, normal contours, and a normal shape to the facial features, her face is attractive despite her scars. The fractional CO₂ laser would greatly further improve her appearance today.

41.4.2 Contour and Proportion

Contractures are the most common result of burn injuries and the tension generated by the contractures alters contour and proportion. Contour is affected by hypertrophic scarring bowstringing across concave surfaces, and compression of the subtle convex and concave surfaces that compose normal human anatomy. Tension, in addition, shifts mobile structures, particularly facial features, in ways that adversely affect normal proportions. The effect of this can be devastating as demonstrated in ► Fig. 41.4. Profound improvement can be accomplished by restoring normal facial proportion and the normal shape of features despite the persistence of extensive facial scarring. These facial proportions are well defined in the literature and can be further studied in the works listed in the “Suggested Readings” section, given that it is beyond the scope of this chapter. Focusing on treating burn scars can be counterproductive. Emphasizing the restoration of normal anatomic proportions and relationships is uniformly rewarding when accomplished. If they are not restored, the patient’s appearance will always be displeasing to the eye, even if the amount of actual physical scarring has been decreased by replacing it by either graft or flap material.

41.4.3 Tension

There is a well-recognized phenomenon of “invisible skin loss” associated with medium second-degree depth or deeper burns resulting in a tightening effect, especially for burns of the face or the dorsum of the hand. Such tension may be obvious when examining deforming contractures of mobile soft tissues such as the eyelids or lips but only subtle in its compression of soft-tissue contours. Relaxed scars are indeed happy scars. This can be easily observed in scars under tension that are erythematous, hypertrophic, pruritic, painful, and tender. Once tension is

relieved, such scars will respond in kind and become flattened, softer, paler, and less symptomatic (► Fig. 41.2 and ► Fig. 41.3). Relief of tension is a practical, achievable, and essential component of operative treatment of burn scar injury.

41.4.4 Donor Sites

Burn scar deformities rarely exist in isolation and are often associated with extensive burns of the patient’s entire body. This presents a major difficulty in burn reconstruction as donor site access may be limited. The principles already set forth allow for intelligent decision-making when considering choice of procedure and the judicious use of available donor sites. This also applies to the choice of reconstructive modality as it relates to either graft or flap variety as that will have an effect on successful outcome. It cannot be overstated that each burn reconstructive case is unique and operative treatment must be individualized.

41.4.5 Aesthetic Units

The concept of aesthetic units has had a profound effect on plastic surgical thinking in reconstructive surgery, especially as it relates to burn reconstruction. It is critical to keep in mind aesthetic units during reconstructive operative treatment for burns; however, it should not overtake common sense. Excision of normal skin is rarely indicated and scar revisions with Z-plasties can allow for profound changes that camouflage scars. The sites of treatment can then be returned to more proportional, well-contoured, and tension-free units that allow for return of form and function to near baseline levels. The elimination of visible scarring is not realistic and such a focused pursuit of an elusive goal can and will lead to unfortunate outcomes and significant iatrogenic trauma.

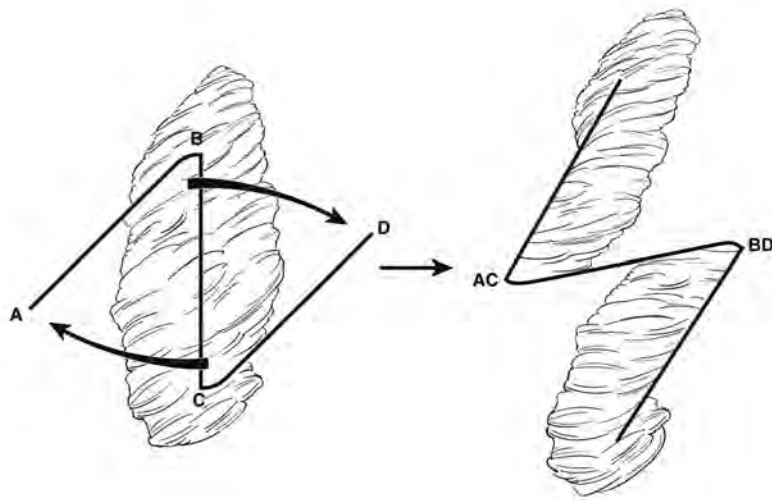


Fig. 41.5 The Z-plasty in burn scars relieves tension by lengthening the scar. It also can be used to narrow the scar, level the scar, and make it less conspicuous by creating irregular borders.

41.4.6 Z-Plasty

The Z-plasty is well described for its ability to lengthen linear scars by recruiting relatively lax adjacent lateral tissue and is an essential and powerful tool in operative burn scar treatment. As mentioned previously, its use on a scar causes a profound, beneficial effect on the physiology of the scar. This is consistent with the principle that scars are living things and not static entities. A properly performed Z-plasty accomplishes two important goals: (1) it decreases longitudinal tension on the scar and (2) it decreases the width of the scarred area. When a properly designed Z-plasty is used, especially a 60-degree limb Z-plasty, this will theoretically lengthen a scar by 75% and narrow it by approximately 30% (► Fig. 41.5). It will also help to camouflage the scar by making its borders more irregular, thereby making it less conspicuous to the human eye. In fact, after a successful Z-plasty, hypertrophic scars resolve rapidly, elasticity improves, and significant scar narrowing is accomplished (► Fig. 41.6). Most importantly, even though grafts and flaps can and do accomplish a similar goal of relieving tension, the use of the Z-plasty does so in a simple, elegant, powerful, and much less morbid and technically difficult fashion. When incising Z-plasty flaps, general principles include that the tips should be cut perpendicular to the central limb for a short distance to add additional tissue to the flap tips and improve blood supply. Also, a Z-plasty should be designed so that once the transposition of the flaps is completed the tight transverse limb of the Z-plasty is located where a normal concavity would occur. Please refer to the patient examples for demonstration of the use of Z-plasty in representative selected cases.

41.4.7 Grafts

It is well established that skin grafts are critical as a tool in burn reconstruction. We have already discussed about considerations with regard to donor site selection, but a few

generalizations will be further helpful as it relates to some additional facets of graft choice and use. Grafts can be divided into split thickness and full thickness, and an entire discussion about the indications and merits of one versus the other is beyond the scope of this chapter. Some generalizations, however, are appropriate to mention. Split-thickness skin grafts contract more, have more propensity to wrinkle, and typically will remain shiny with a “glossy finish” look to them. Split-thickness skin grafts are usually the best alternative in many different areas of the body to relieve broad areas of contracture that are not suitable for reconstruction with Z-plasty or local flap tissue rearrangement. Even on the face, when used in inconspicuous locations, split-thickness grafts can decrease tension on hypertrophic scarring and enhance scar maturation (► Fig. 41.7). Caution must be used in split-thickness skin grafting in dark-skinned patients because hyperpigmentation is a problem that is encountered frequently. Full-thickness skin grafts contract less, have a matte finish, often retain skin appendages, and are therefore the ideal skin replacement if the recipient bed is adequate and the price of the donor site is worth the potential improvement. Full-thickness skin grafts are reliable “workhorses” for facial burn reconstruction, particularly when resurfacing broad, conspicuous areas of the face in aesthetic units (► Fig. 41.8). Unlike flaps, over time these grafts allow for excellent facial expression. Treatment with the fractional CO₂ laser makes the grafts feel thinner and more elastic, further improving their advantages over flaps on the face. When using full-thickness grafts for facial reconstruction, all contractures should be eliminated, the defect overcorrected, and larger grafts than one might think necessary must be used. Normal skin should never be excised in burn patients as the junctional suture lines are easily revised with Z-plasties and laser therapy, as demonstrated in this patient, and the fundamental problem is a skin deficiency. Additionally, postoperative management with conformers and pressure therapy is particularly essential when using these grafts.

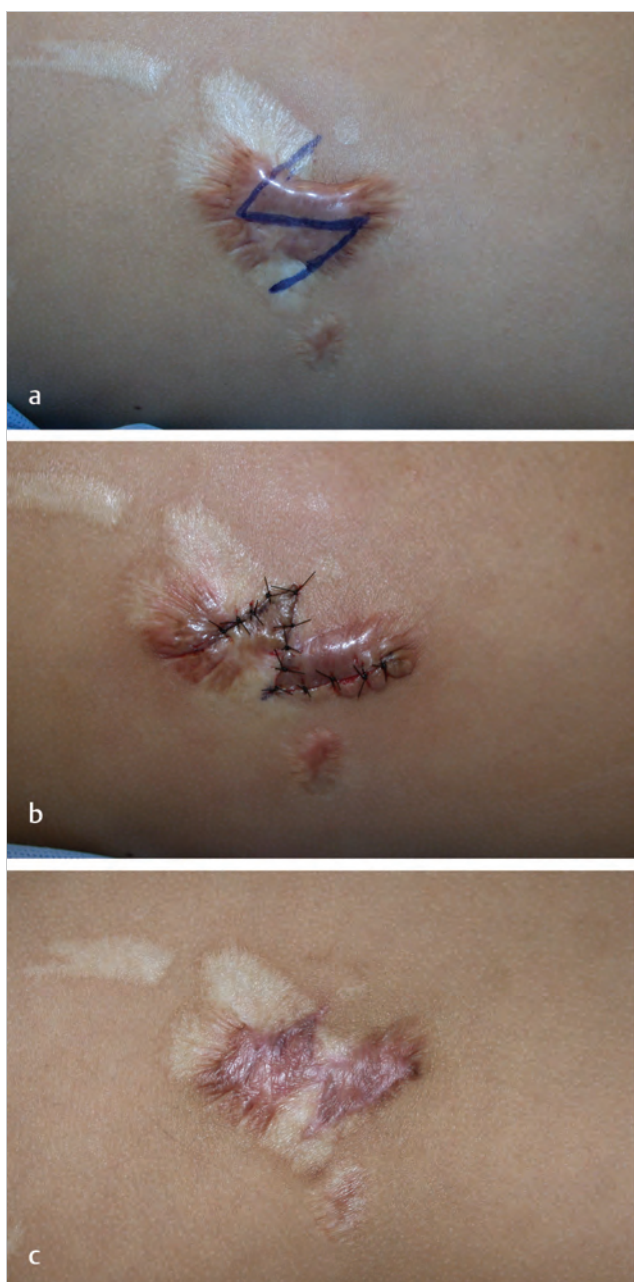


Fig. 41.6 (a) Preoperative: This hypertrophic, contracted scar is raised, erythematous, shiny, and pruritic. (b) Postoperative: After the Z-plasty has been incised and transposed, the scar is longer, completely level with the adjacent skin in the center, and irregular in outline. (c) Three weeks postoperation, the change in the scar is profound. Flattening is continuing, erythema is resolving, and pruritus is significantly less. Changing a scar's environment changes its behavior.

41.4.8 Flaps

Flaps must be judiciously and skillfully used, whether with or without tissue expansion. There can be problems and iatrogenic deformities created when flaps are performed contrary to the principles laid forth previously. In particular, flaps used in the facial region are particularly prone to misuse. This is because

the thickness of skin flaps from almost all donor sites is greater than that of normal facial skin and will tend to mask normal facial movement and expression. The same is true regarding tissue expansion and flap design and use in the lower extremities. Conversely, tissue expansion and flap usage for burn scar scalp alopecia is an ideal treatment modality. It is also important to realize that the size of flaps required to adequately relieve contractures can be remarkably large and this can easily create extrinsic contractures that can adversely affect form and function. Inadequate contracture releases with flaps are common, and iatrogenic deformities from flap usage are unfortunately very common (► Fig. 41.9). In addition, color and texture match are very important considerations in flap design, and donor sites must be carefully chosen with this in mind. However, despite these cautionary points, well-chosen and expertly performed flaps can be very helpful in selected cases and are essential for complex injuries such as electrical burns.

41.4.9 Lasers

Advances in laser therapy have greatly facilitated scar treatment and rehabilitation, further decreasing the indications for scar and, in particular, burn scar excision. Laser scar therapy represents a well-established, although newer, technology that is not only promising, but also still remarkably underutilized in the multidisciplinary treatment of burn scars and other traumatic scars. This has heralded changes to existing scar treatment paradigms that allow for integration of lasers as well as combination therapies into future scar treatment protocols. Pulsed dye laser, and now fractional photothermolysis, therapies have emerged over the past decade as successful alternatives to excision in patients with hypertrophic burn scars. Lasers have been proven to be safe, consistently effective, and demonstrate potential functional as well as cosmetic benefits in burn scar treatment. The basic premise is that laser treatment of scars allows for the production of microscopic patterns of thermal injury in the dermis, which stimulates the complex process of tissue remodeling. Laser treatment is particularly effective when combined with Z-plasty release or some of the other modalities mentioned earlier (► Fig. 41.3). Prospective, randomized trials are in progress to evaluate how to optimize already clearly effective laser treatment protocols and will provide further insights on how to take maximum advantage of this powerful tool in the plastic surgical armamentarium.

41.5 Complications

As has been mentioned thus far, each of the considerations discussed has also been tempered with caution with regard to unfavorable results and complications, especially iatrogenic, that may occur even with the best intentions and most precisely crafted nonoperative management or operative treatment plans. Often, many of the complications seen and the secondary “deformities” created by nonprincipled treatment plans are thankfully avoidable or able to be minimized such that forward progress is maintained. Frequently, Z-plasty repair may lead to tip necrosis or wound separation that can both be



Fig. 41.7 (a) This 3-year-old male sustained severe burn injury to the head and neck in a house fire in China. The facial burns healed without grafting. He was adopted and brought to the United States. **(b)** Eight years later, after Z-plasties, small releases/grafts, and treatment with the pulse dye laser and fractional CO₂ laser, his facial scars are completely rehabilitated. No scars were excised.

treated with local dressing care until re-epithelialization ensues. Typically, in these cases, because tension has been relieved by the Z-plasty, wound healing goes on to be quite favorable and without hypertrophic sequelae. Grafts and flaps have their own unique considerations, and the informed consent process must focus on bleeding, infection, partial or complete graft or flap loss, unfavorable scarring, tissue expander infection or extrusion, donor site complications, and need for further procedures due to secondary iatrogenic deformities created by primary operative intervention.

As it relates to laser therapy, scars seem to tolerate this well with a remarkably low rate of complication. Immediate onset of transient erythema and localized swelling are well-known issues, as well as pinpoint bleeding, depending on laser settings and depth of penetration. Reports of infection and new or worsening scarring do exist in the literature but are very infrequent, and overall the safety profile is exceptional. More important, however, are the safety considerations around laser therapy for not only the patient, but also the treatment provider and team. Eye protection, fire safety protection, smoke evacuation, and other safety measures are critical and must be closely adhered to whenever using any laser device.

41.6 Conclusion

Pediatric burn care and reconstruction have witnessed significant and remarkable advances in the care of acute and secondary burn patients. Owing to typical child's robust health, robust nutritional status, and lack of comorbidities at the time of most burn injuries, even those unfortunate patients with greater than 90% total body surface area burns are surviving and benefiting from the expertise and multidisciplinary care provided to them. Therefore, once the acute burn chaos has

subsided, successful secondary pediatric burn scar reconstruction can restore these patients sufficiently that they may enjoy a happy, healthy, and very productive life. This is evidenced by well-reported, large series in the literature that demonstrate excellent long-term outcomes when compared with normal controls. What is most important is that the essential qualities to ensure such outcomes are adhered to: patience, persistence, determination, adherence to fundamental principles, and a curiosity to keep an open mind as newer technologies become available. Thoughtful and skillful application of these qualities to the treatment of pediatric burn injuries will lead to optimization of outcomes and provide gratification to the patients and providers of care alike. The critical principle of pediatric burn reconstruction is learning that has been espoused thus far in this chapter, namely, to understand, appreciate, and most favorably influence the process of wound healing and scar maturation. This is a goal that is attainable and one that we must steadfastly strive to achieve for each and every patient we as pediatric plastic and reconstructive surgeons have the privilege to care for.

41.7 Key Points

- Scars are your friends.
- Relaxed scars are happy scars.
- Scar excision is an oxymoron.
- Adhere to fundamental principles and do not rely on specific techniques as techniques are transient, but principles stand the test of time.
- Laser therapy should be included as early as possible in the care of burn scar injuries and considered as another critical tool in the armamentarium of burn care and treatment.



Fig. 41.8 (a) An 8-year-old female who sustained 65% total body surface area flame burns in a house fire. Her facial split-thickness grafts were hyperpigmented, contracted, irregular, and conspicuous. (b) At age 11, the split-thickness skin grafts were replaced with a full-thickness graft from her chest, the best available match for color and texture. The donor site was closed with a split-thickness graft. (c) Only the most noticeable area of the split-thickness skin graft was excised. The surrounding tissue was released to allow it to retract. A much larger graft than the measured defect was harvested. (d) Multiple scar revisions with Z-plasties at the margins of the full-thickness skin graft and treatment with the fractional CO₂ laser resulted in significant improvement and a happy patient.



Fig. 41.9 (a) This 12-year-old male has a normal facial appearance as well as hypertrophic burn scars with minimal contractures. Today, the scars could easily be rehabilitated with Z-plasties and fractional CO₂ laser therapy. (b) Scar excision was performed and cheek resurfacing carried out with a tissue expanded cervicopectoral flap. (c) Flap tip ischemia and downward tension are frequent complications. (d) Following excision of scar and flap resurfacing, there is lower lid displacement, malar flattening, loss of jawline definition, and increased contracture of the oral commissure and lips.

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42 Melanocytic Nevus

Aladdin H. Hassanein and Arin K. Greene

Summary

Melanocytic nevi are common in children. The primary indication for removal is to improve a deformity. Giant nevi are associated with a low risk of malignant degeneration. Generally, serial excision is preferred to skin grafting or tissue expansion when feasible.

Keywords: melanocytic, nevus, melanoma, pediatric

42.1 Introduction

Melanocytic nevi are one of the most common pediatric cutaneous conditions; lesions can be present at birth or acquired. Congenital nevi affect 1% of the population and form when nevomelanocytes become ectopically located during migration from the neural crest to the epidermis. Congenital melanocytic nevi can cause a deformity and psychosocial morbidity. Patients and families frequently are anxious about malignant degeneration, but the risk of transformation to melanoma often has been overestimated. This chapter will focus on evidence-based management of congenital melanocytic nevi.

42.2 Diagnosis

Melanocytic nevi are diagnosed by history and physical examination. Age of onset (congenital or acquired), asymmetry, border irregularity, color, size, and symptoms should be elicited. In contrast to acquired nevi, histopathology of congenital lesions exhibits nevomelanocytes in the lower two-thirds of the dermis and subcutis involving sebaceous glands, hair follicles, vessels, and nerves. Congenital melanocytic nevi can be classified based on their diameter or percent total body surface area (TBSA). Although a consensus definition does not exist, lesions usually are categorized as small (<1.5 cm), medium (1.5–10 cm), large (11–20 cm), and giant (>20 cm or >2% TBSA). We favor the definition of giant nevi greater than 2% TBSA because it is easy to assess clinically by determining if the lesion is twice the area of the infant's hand (1% TBSA). Giant congenital melanocytic nevi are rare (1/20,000 births) but problematic.

Imaging is unnecessary to diagnose congenital melanocytic nevi. However, patients with giant lesions are at risk for melanocytes involving the leptomeninges. Neurocutaneous melanosis can cause developmental delay, hydrocephalus, and seizures. Children with lesions located in the midline head or trunk and those with multiple satellite nevi are at greatest risk for neurocutaneous melanosis. Magnetic resonance imaging of the central nervous system is performed during infancy to rule out neurocutaneous melanosis in patients with giant pigmented nevi.

Biopsy of a congenital melanocytic nevus is indicated to exclude melanoma if there is a change in appearance, ulceration, bleeding, or enlargement. Additional criteria include the "ABCDs": Asymmetric shape, Borders are irregular, multiple Colors are present, and the Diameter is greater than 6 mm. Malignant degeneration is very rare (0.03% lifetime risk) for

small nevi. The rate of melanoma for giant lesions historically has been overestimated (up to 12%). However, recent studies suggest that malignant degeneration is approximately 3%. The incidence of melanoma is greatest within the first 3 years of life, followed by another peak during adolescence. Nevi at highest risk for malignancy are those located on the trunk and lesions covering an extensive area. Melanoma has an approximately equal chance of occurring in the main nevus, a satellite lesion, or in the central nervous system.

42.3 Nonoperative Management

Congenital melanocytic nevi can be observed and do not mandate intervention. Nonexcisional treatments (e.g., curettage, dermabrasion, hydroquinone, laser) may be used to improve the appearance of the nevi. These nonextirpative methods are reserved for lesions in aesthetically sensitive areas where resection would cause greater disfigurement than the nevus. However, the cancer risk is not eliminated and fibrosis may complicate later histopathology.

42.4 Operative Management

Indications for removing congenital melanocytic nevi are (1) concern for malignancy, (2) lesions causing psychosocial morbidity, or (3) to reduce the risk of malignant transformation for giant congenital nevi.

42.4.1 Timing of Resection

Concern for melanoma requires immediate excision of the suspicious area. Removal of a lesion that is likely to cause psychosocial morbidity can be delayed until 3 to 4 years of age when self-esteem and long-term memory develop. Extirpation of nevi on the scalp during infancy is advantageous because the tissue is particularly lax during this period. Lower extremity lesions should be removed in infancy while the patient is nonambulatory to reduce the risk of suture line dehiscence.

Because the peak incidence of malignant conversion for giant lesions is greatest within the first 3 years of life, prophylactic resection to decrease the occurrence of melanoma is most effective if completed before this time. We begin excision at 6 months of age when we are trying to completely remove a giant nevus to reduce the likelihood of malignant conversion. We avoid elective procedures prior to 6 months of age because the risk of anesthesia is higher.

42.4.2 Small and Medium Congenital Melanocytic Nevus

Small and medium-sized lesions are resected to aesthetically improve a deformity or if the clinical appearance becomes worrisome for malignancy. These nevi usually can be removed leniently and closed linearly in one stage. Circular excision and

purse-string closure may be performed for small, round nevi on the face to minimize the length of the scar; lenticular excision of a round lesion results in a scar two to three times its diameter (► Fig. 42.1). Approximately 50% of patients are satisfied with the circular cicatrix, which can appear as an acne or chicken pox scar. One-half of patients undergo a second stage to convert the circular scar into a line that is approximately the same length as the diameter of the original lesion. Lesions

located in difficult locations can result in more challenging reconstructive problems (e.g., nose, eyebrow; ► Fig. 42.2, ► Fig. 42.3, ► Fig. 42.4, ► Fig. 42.5). Raised dermal nevi that are located in unfavorable locations (particularly the nose) can be improved by shave excision to the papillary dermis (► Fig. 42.6). The patient's appearance and contour will be improved without a scar, but the individual likely will require repeat shave excision in the future if the nevus re-enlarges.



Fig. 42.1 Small nevus of the chin causing lowered self-esteem treated with circular excision and purse-string closure to limit the length of the scar. (a) Preoperative view. Note the marking for lenticular resection and increased size of scar if this method was chosen. (b) After circular excision with approximately 1-mm margins. (c) One year postoperatively. The patient was pleased with the appearance of the scar and did not want a second procedure to convert the circular scar into a line.



Fig. 42.2 A child with a small nevus treated with excision and cheek advancement to place the scar in the alar crease. (a) Preoperative view. (b) Markings prior to resection. (c) After removal. (d) Postoperative image.



Fig. 42.3 Infant with a medium-sized nevus in an unfavorable location. It was treated with dermabrasion, hydroquinone, and serial excision to avoid skin grafting or a local/regional flap reconstruction. (a) Preoperative appearance. (b) Intraoperative view following dermabrasion. (c) After dermabrasion, hydroquinone, and an intermediate stage of serial excision. (d) Final appearance after completing serial excision. Note the patient ultimately had a single linear skin at the junction of aesthetic subunits.

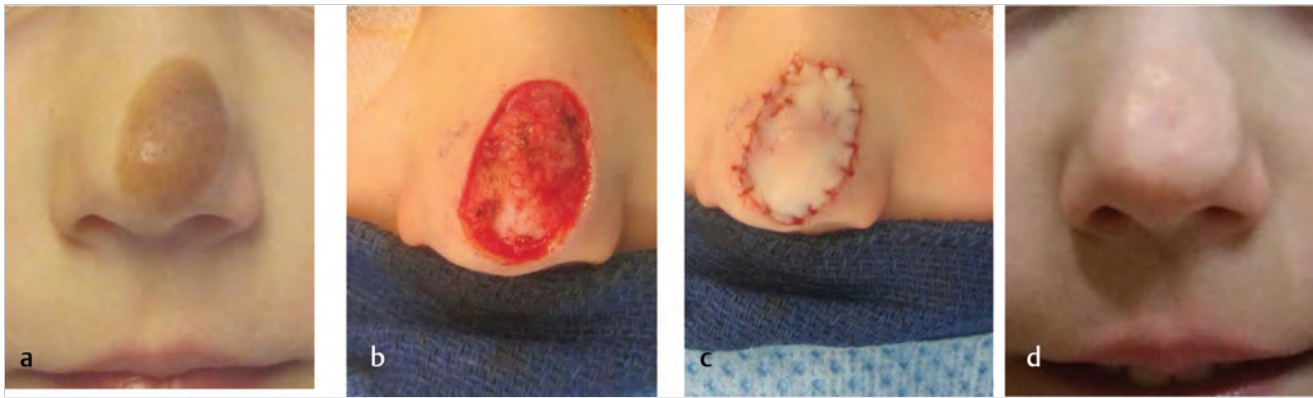


Fig. 42.4 A child with a medium-sized nevus on the nose reconstructed with skin grafting. (a) Preoperative view. (b,c) Following resection and reconstruction with a full-thickness retroauricular skin graft. (d) One year postoperatively.

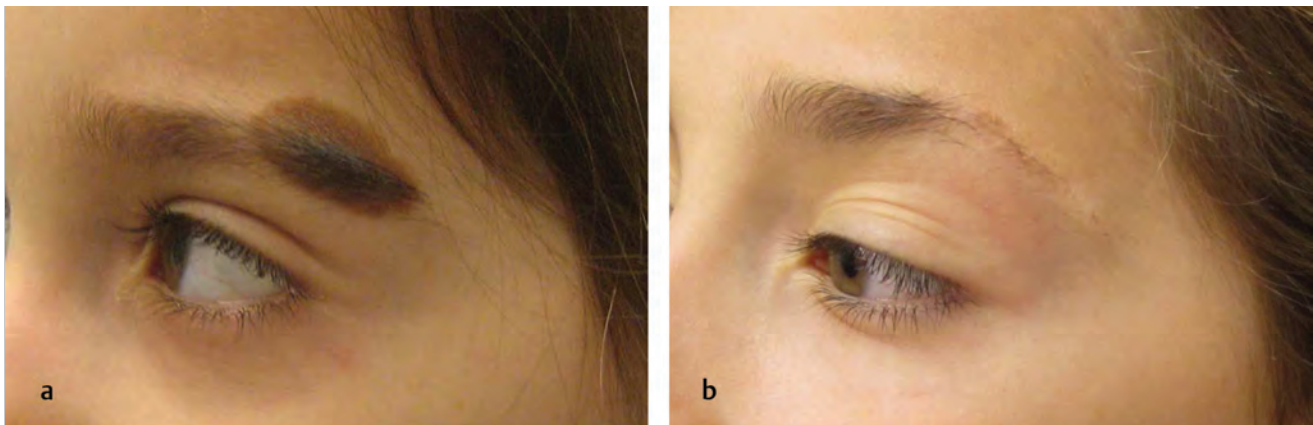


Fig. 42.5 An adolescent patient with a medium-sized nevus involving the eyebrow managed with lenticular excision. (a) Preoperative appearance. (b) Postoperative view.



Fig. 42.6 Patient with a raised dermal nevus of the nasal tip managed with shave excision.

Small/medium melanocytic nevi on the face being removed for “cosmetic” reasons should have minimal gross margins (0–1 mm) to ensure the most favorable scar. If no atypical cells are present, then positive margins require no further treatment, although there is a low risk (~1%) of minor re-pigmentation along the scar, which can be resected secondarily. If histopathology shows cellular atypia then re-excision to negative margins is required. Small/medium nevi located in nonaesthetically important areas or those at risk for atypia can be extirpated with 1- to 2-mm margins.

42.4.3 Large/Giant Congenital Melanocytic Nevi

Although giant lesions are at greater risk for melanoma, generally large and giant nevi are approached similarly. These lesions can be unsightly, but complete extirpation may create a greater deformity. We agree with Zuker and colleagues that the risk of malignancy should be weighed against the aesthetic outcome. The frequency of malignant conversion (~3%) for giant nevi is lower than historical estimations; no evidence proves that the frequency of melanoma is decreased by prophylactic excision. Therefore, we typically manage lesions nonoperatively if removal would be more disfiguring or compromise function. Giant nevi that are not removed are monitored by a dermatologist; biopsies are performed if an area becomes suspicious for malignancy.

Techniques to remove giant congenital melanocytic nevi are (1) serial excision, (2) skin grafting, and (3) tissue expansion. We prefer serial excision because it has many advantages compared to skin grafting or tissue expansion. First, the method is simpler and only necessitates wide skin undermining to close the wound. Second, the operations are short and performed on an outpatient basis. Third, patients have a quicker recovery and less risk of operative morbidity. Fourth, complications (e.g., infection, dehiscence) do not adversely affect the final outcome. A dehiscent wound can be allowed to heal secondarily and the wider scar excised during the next stage. If dehiscence or infection complicates skin grafting or tissue expansion, the

consequences are more problematic. Finally, children are left with a single linear scar, which is aesthetically more favorable than skin grafting or tissue expansion. A limitation of serial excision is that it relies on the ability to recruit adjacent tissue using subcutaneous undermining; it is unable to be performed in circumferential extremity lesions and areas that lack skin laxity. Several stages may be required; we have performed as many as five excisions for extensive lesions.

Skin grafting following resection of giant congenital melanocytic nevi allows for treatment in one procedure (► Fig. 42.7). However, the recipient site appears as an indented, hairless patch with a color distinct from the surrounding native skin. Graft failure can occur and a painful donor site scar is created. Children usually require hospital admission to ensure revascularization of the graft. Patients are limited postoperatively to ensure immobilization of the area.

Tissue expansion also is used to treat giant congenital nevi. Similar to serial excision, tissue expansion replaces the lesion with adjacent, local tissue, which is better aesthetically than a skin graft (► Fig. 42.8). However, the technique requires a procedure to place the expander, multiple outpatient visits for filling, and an additional operation to remove the implant, excise the lesion, and inset the expanded skin. Rotation, transposition, and/or advancement of expanded flaps create longer and more numerous scars compared to a single line from serial excision. As the volume in the expander increases and becomes more visible to others, patients can experience pain and psychosocial morbidity. There is a risk for expander infection, malposition, deflation, malfunction, and extrusion. Tissue expansion is most problematic in the pediatric population.

42.4.4 Head/Neck

Melanocytic nevi involving the head/neck are more likely to undergo resection because they cause an obvious deformity. Large lesions over the scalp, forehead, periorbital area, nose, and ear are problematic to treat with serial excision because of poor skin laxity and difficulty recruiting local tissue with subcutaneous undermining (► Fig. 42.9, ► Fig. 42.10, ► Fig. 42.11).

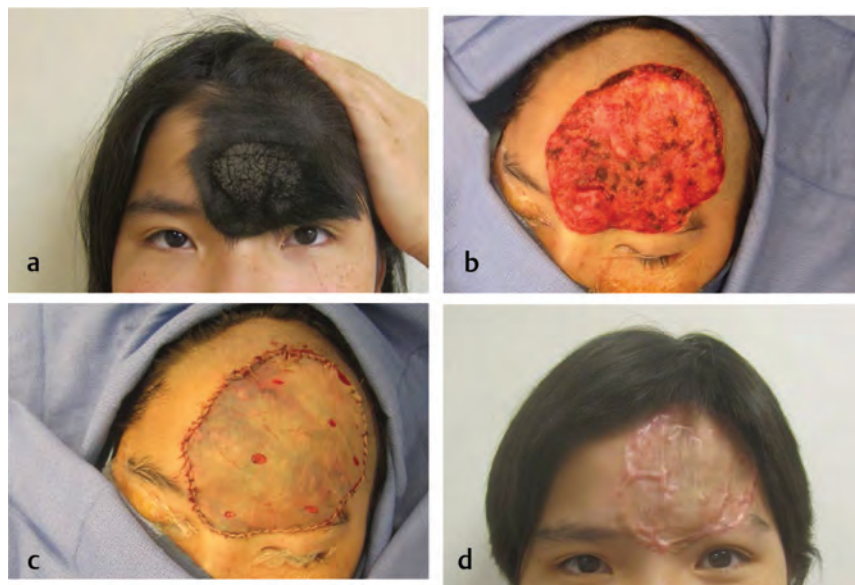


Fig. 42.7 An adolescent patient with a giant congenital melanocytic nevus of the forehead and scalp managed with skin grafting. (a) Pre-operative photo. (b) Following resection. (c) Reconstruction with a split-thickness skin graft. (d) Three months postoperatively.



Fig. 42.8 Giant congenital melanocytic nevus of the scalp and forehead treated with tissue expansion. (a) Prior to resection. (b,c) Following tissue expander placement and filling. (d) After removal of the expander and flap advancement. (e) Postoperative image.



Fig. 42.9 Medium-sized congenital melanocytic nevus of the cheek treated with serial excision. (a) Preoperative image. (b) After first stage removal. (c) Following second resection.

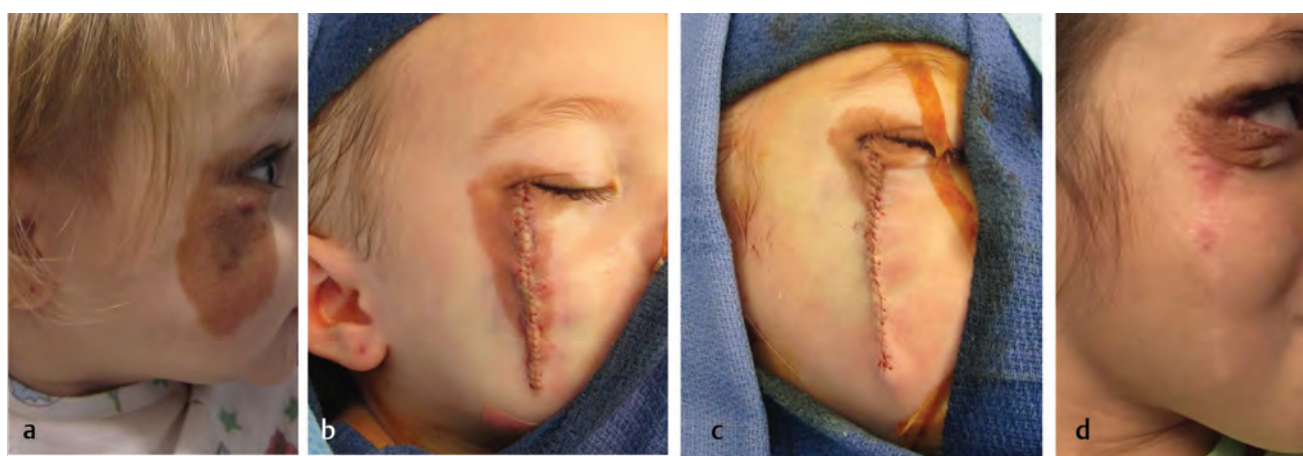


Fig. 42.10 Serial excision of congenital melanocytic nevus of the cheek and eyelids. (a) Initial presentation. (b) Following first resection. (c) After intermediate stage. (d) Postoperative appearance following serial excision. Note that the eyelid is not amenable to serial removal and will be reconstructed with full-thickness skin grafts.

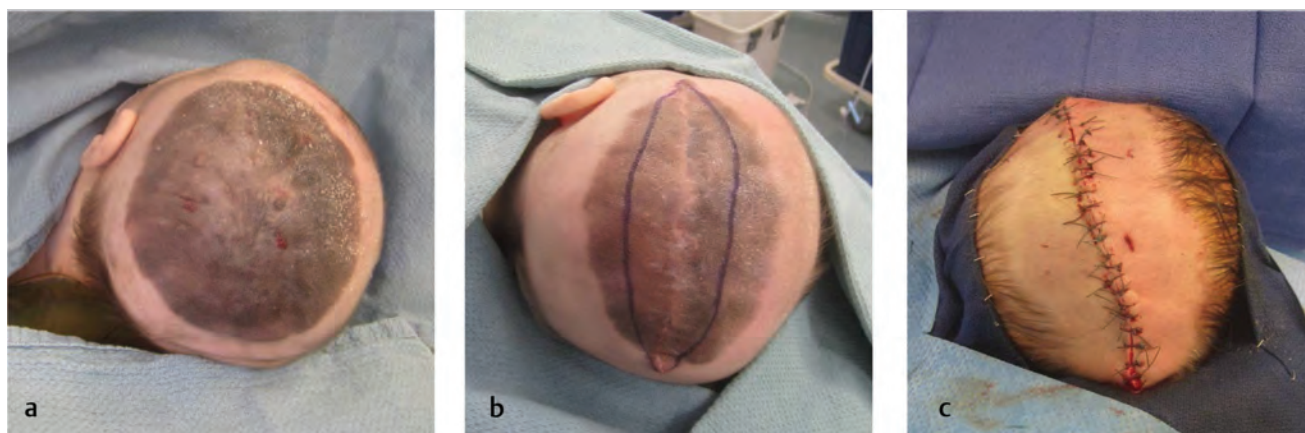


Fig. 42.11 Giant congenital melanocytic nevus of the scalp managed with serial excision. The procedure was initiated at 3 months of age to take advantage of infant scalp laxity. (a) Preoperative image. (b) Prior to second-stage resection. (c) Postoperative view after the final fifth excision.

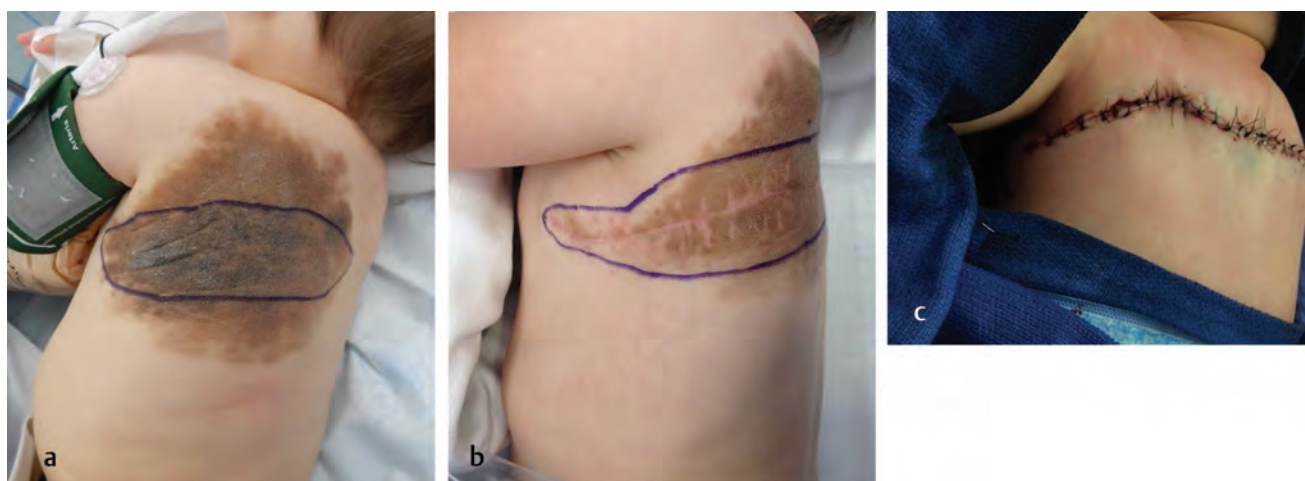


Fig. 42.12 A child with a giant congenital nevus of the trunk. (a) Preoperative view. (b) Prior to second-stage resection. (c) After the fourth and final stage.

Tissue expansion is preferred for scalp lesions that cannot be removed with serial excision. Large forehead nevi with a paucity of normal skin to expand/recruit can be treated with skin grafting. Periorbital lesions often require skin grafting. The benefits of removing a congenital nevus of the head/neck must justify creating an unaesthetic, iatrogenic deformity that may result from resection (e.g., facial nerve injury during tissue expansion).

42.4.5 Trunk

Serial excision is our preferred technique for giant nevi of the trunk. Some authors favor tissue expansion to treat nevi on this region, but we have found that most lesions are amenable to removal with serial excision (► Fig. 42.12). Serial excision is lower on the reconstructive ladder and results in a single, linear scar. Although more stages may be required with serial excision compared to skin grafting or tissue expansion, avoiding the complications and potential morbidity warrants the additional procedures. We prefer to observe patients with “bathing-suit” nevi involving the entire trunk because the nevus is hidden by

clothing (► Fig. 42.13). We do not believe the risks and benefits of extensive skin grafting and/or tissue expansion are favorable for these patients.

42.4.6 Extremity

Congenital melanocytic nevi of the extremities have lower risk of malignant transformation than other areas. In addition, these lesions usually can be concealed by clothes. Most large/giant nevi of the limbs are amenable to treatment with serial excision (► Fig. 42.14). However, circumferential lesions are unable to be removed with serial excision because adjacent, normal tissue cannot be recruited. Therefore, skin grafting or tissue expansion is required. Large hand and foot nevi also are difficult to treat with serial excision. Skin grafting of hand lesions can result in contracture and affect function. Tissue expanders often become infected in the lower extremities. Thus, the risk/benefit profile of treating extremity lesions must be carefully considered. Large circumferential lesions, particularly involving joints, can be resected and reconstructed with full-thickness skin grafts that have been expanded from the lower trunk.



Fig. 42.13 Giant congenital melanocytic nevus involving the trunk and lower extremities. The lesion is hidden by clothing and was managed with observation by a dermatologist.

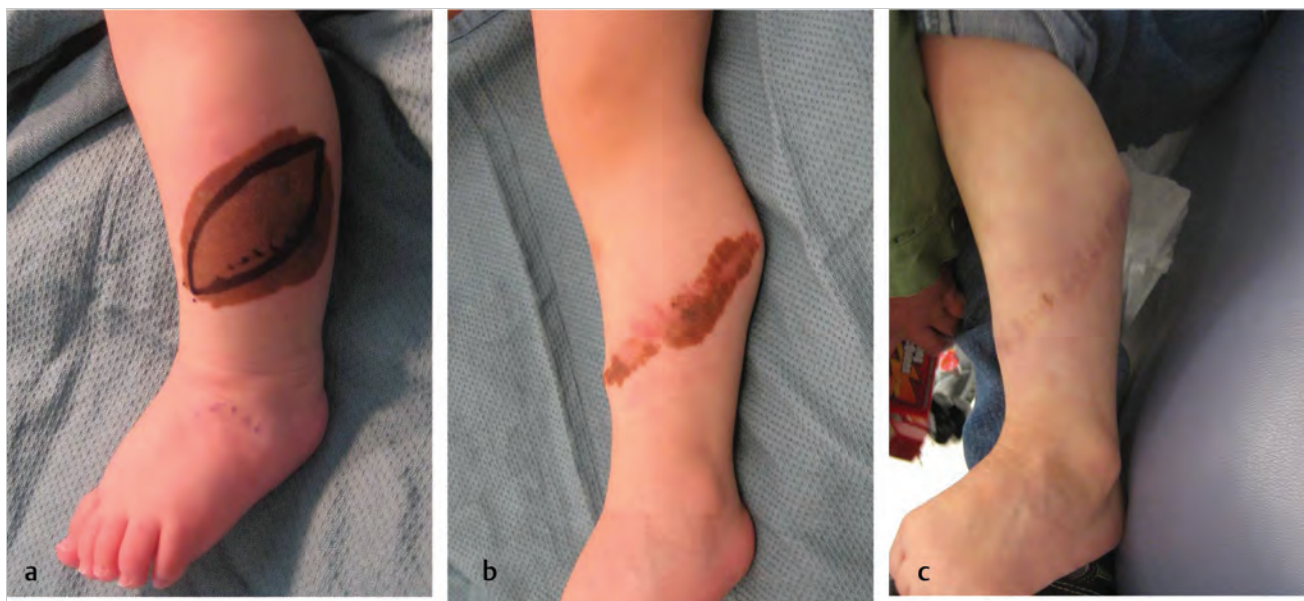


Fig. 42.14 Large congenital melanocytic nevus of the leg treated with serial excision. (a) Preoperative image. (b) Prior to third and final resection. (c) Postoperative appearance.

42.5 Complications

The greatest concern in managing congenital melanocytic nevi is a delay in diagnosis of melanoma. Lesions that are treated nonoperatively should be followed serially by a dermatologist and biopsied if there is a suspicious change in appearance. Patients with a giant congenital melanocytic nevus are assessed with magnetic resonance imaging for neurocutaneous melanosis in order to anticipate possible developmental delay, hydrocephalus, and seizures. Complications of serial excision include infection and wound dehiscence. Skin grafting can result in partial or complete failure and cause a donor site scar. Tissue expansion is associated with many difficulties and is more challenging than serial excision or skin grafting. There is a risk for expander infection, malposition, deflation, and extrusion. These

complications may limit the amount of available donor tissue or cause abandonment of this reconstructive option.

42.6 Conclusion

Congenital melanocytic nevi are common pediatric skin lesions that affect 1% of the population. They cause a deformity and psychosocial morbidity. Small and medium-sized lesions can be removed for aesthetic improvement or if they are suspicious for malignancy. Giant congenital melanocytic nevi are at risk for neurocutaneous melanosis and malignant transformation. These lesions can be resected using serial excision, skin grafts, or tissue expansion. The benefits of removal should be weighed against the morbidity of extirpation.

42.7 Key Points

- Congenital melanocytic lesions are common and are categorized as small, medium, large, or giant.
- Removal is performed for aesthetic improvement or if there is a concern for malignancy.
- Giant congenital melanocytic nevi have a low risk for malignant transformation (~3%).
- Resection is not mandatory; lesions can be observed and followed for suspicious changes.
- Extirpation of giant lesions must be weighed against the morbidity of the procedures and reconstruction.

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43 Pediatric Skin Lesions

Aladdin H. Hassanein and Arin K. Greene

Summary

Cutaneous lesions are common in children. Although almost always benign, they may cause psychosocial morbidity and anxiety concerning malignant degeneration. This chapter reviews the diagnosis and management of the most frequently treated pediatric skin lesions: acrochordon, dermoid cyst, epidermal cyst, epidermal nevus, granuloma annulare, hypertrophic scar/keloid, lipoma, juvenile xanthogranuloma, mucocele, nevus sebaceous, neurofibroma, pilar cyst, pilomatrixoma, Spitz nevus, and verruca vulgaris.

Keywords: dermoid, cyst, keloid, lipoma, mucocele, nevus, neurofibroma, pilomatrixoma, skin lesion

43.1 Introduction

Cutaneous lesions are common in children and frequently are managed by pediatric plastic surgeons. They are almost always benign, although some can be premalignant. Skin lesions may cause a deformity and psychosocial morbidity. Patients and families often are anxious about malignant degeneration. Most conditions are diagnosed by history and physical examination;

imaging or biopsy rarely is indicated. Lesions necessitating removal usually require a general anesthetic in young children; adolescents may tolerate excision using local anesthesia. This chapter reviews the most common pediatric skin conditions that are encountered by plastic surgeons: acrochordon, dermoid cyst, epidermal cyst, epidermal nevus, granuloma annulare hypertrophic scar/keloid, juvenile xanthogranuloma, lipoma, nevus sebaceous, neurofibroma, mucocele, pilar cyst, pilomatrixoma, Spitz nevus, and verruca vulgaris (► Table 43.1).

43.2 Diagnosis

Most cutaneous lesions can be diagnosed by history and physical examination. Age of onset (congenital or acquired), changes in appearance, and symptoms (e.g., bleeding, infection, pain, ulceration) are elicited. Examination should assess color, location, mobility, size, symmetry, and tenderness. Pinching the lesion in relation to the surrounding soft tissue can determine the depth of involvement. If the skin can be approximated over the lesion, then the abnormality is located beneath the integument (e.g., dermoid cyst, lipoma, etc.).

Imaging rarely is indicated if (1) the diagnosis is uncertain despite clinical evaluation and (2) the results of the study will

Table 43.1 Common pediatric skin lesions

Lesion	Clinical features	Histopathology
Acrochordon	Soft, flesh-colored fibroepithelial polyp	Epidermal hyperplasia; ±adipose central core
Dermoid cyst	Congenital; location in brow, nasoglabellar, or orbital areas	Epithelial lining, pilosebaceous structures in the cyst wall
Epidermal cyst	Proliferation of epidermal cells within the dermis	Lined with epithelium and filled with keratin
Epidermal nevus	Hyperkeratotic lesion	Epidermal granular layer, increased basal layer pigment, inflammatory dermal infiltrate
Granuloma annulare	Inflammatory condition; ringlike, erythematous lesion	Degenerative collagen surrounded by histiocytes, fibroblasts, and lymphocytes
Juvenile xanthogranuloma	Pinkish-yellow; can regress	Histiocytes and giant cells
Keloid	Grow beyond the wound borders; predilection for chest, ear, shoulder	Fibrous tissue, and irregularly arranged dermal collagen
Lipoma	Subcutaneous, mobile lesion with normal overlying skin	Benign adipose overgrowth
Mucocele	Oral pseudocyst from trauma or obstruction of a minor salivary gland	Mucin-fluid, no epithelial lining
Nevus sebaceous	Yellow-orange hairless plaque; predilection for the scalp and face	Cutaneous hyperplasia of adnexal elements
Neurofibroma	Benign peripheral nerve sheath tumor	Fibroblasts, mast cells, and Schwann cells
Pilar cyst	Subcutaneous hair follicle outer root sheath cyst that occurs on the scalp	Keratin filled; wall of stratified squamous epithelium
Pilomatrixoma	Firm, fixed mass with a bluish hue; can ulcerate	Benign calcifying tumor of the hair follicle matrix cells
Spitz nevus	Acquired dome-shaped pink nodule, typically <1 cm	Benign tumor of spindled and epithelioid melanocytes
Verruca vulgaris	Flesh-colored, firm papules	Hyperkeratosis, acanthosis

influence management. Generally, ultrasonography is the first-line imaging modality. It can distinguish solid from cystic lesions and assess vascularity. Magnetic resonance imaging (MRI) or computed tomography (CT) is considered if ultrasonography is unclear. Biopsy to elucidate a histopathological diagnosis prior to extirpation usually is unnecessary. If removal is required, definitive diagnosis will be confirmed with an excisional biopsy.

43.3 Nonoperative Management

Asymptomatic lesions with a low malignant potential can be observed and followed serially. Nonexcisional treatments (e.g., dermabrasion, laser) may be used to improve the appearance of some cutaneous conditions, but the cancer risk is not eliminated and fibrosis may complicate later histopathology.

43.4 Operative Management

Skin lesions are excised because of (1) symptoms, (2) malignant risk, or (3) aesthetic concerns. Removal of a lesion that is likely to cause psychosocial morbidity can be considered between 3 and 4 years of age when self-esteem and long-term memory develop. Most cutaneous disorders that necessitate extirpation are lenticularly excised and closed linearly. Round facial lesions may benefit from circular excision and purse-string closure to give the shortest possible scar. A second stage can be performed to convert the circular scar into a line. Subcutaneous lesions are removed using an incision (or small skin excision) over the abnormality. Disorders beneath the skin usually can be resected with a scar that is shorter than the diameter of the lesion.

43.4.1 Acrochordon

Acrochordon (i.e., skin tag) is a common pedunculated fibroepithelial polyp. It exhibits epidermal hyperplasia; large lesions may have an adipose component. Acrochordons are soft, flesh-

colored, and can be sessile or pedunculated. They have a predisposition to develop in skin folds (e.g., neck, axilla, groin) and occur more frequently in diabetic or pregnant patients. Large lesions can cause discomfort and deformity. Symptomatic acrochordons are usually treated by excision; cryotherapy is a less common option.

43.4.2 Dermoid Cyst

Dermoid cysts are congenital and usually located on the face or cranium. Lesions contain ectodermal and endodermal tissue; lining consists of keratinizing squamous epithelium and pilosebaceous structures. Orbitofacial dermoids are caused by dermal and epidermal cell displacement along embryonic lines of fusion. They are asymptomatic, slow growing, and most frequently located in the frontotemporal, nasoglabellar, or orbital area (the most common site is the lateral brow; ► Fig. 43.1). If a dermoid is suspected, imaging is unnecessary unless the lesion is along the nasal midline. MRI is obtained to determine whether a midline nasal dermoid extends intracranially along a tract. Dermoids located over the nasal bones have a low risk of intracranial extension, while lesions involving the nasal tip have an approximately 50% risk of intracranial communication. Brow and orbital dermoids usually can be removed through an eyelid incision. Nasoglabellar dermoids without intracranial involvement can be excised directly or using an open-rhinoplasty approach. Dermoids with intracranial extension may require a nasal and coronal incision with a craniotomy to remove the entire tract.

43.4.3 Epidermal Cyst

Epidermal (“sebaceous”) cysts are benign proliferations of epidermal cells within the dermis that can occur congenitally or by implantation of epithelial cells into dermis (► Fig. 43.2). Lesions are filled with keratin and lined with stratified squamous epithelium, which contains a granular layer. Epidermal cysts appear as round nodules often with a central pore. They slowly



Fig. 43.1 (a) A child with a dermoid cyst of the lateral brow. (b) An infant with a nasal dermoid. A midline nasoglabellar dermoid requires magnetic resonance imaging to assess for intracranial extension.



Fig. 43.2 A child with epidermal cyst of cheek. Note the round subcutaneous appearance.



Fig. 43.3 A 13-year-old male with a pigmented and hyperkeratotic epidermal nevus of the neck.

enlarge over time, may become infected, and can cause a deformity. Imaging is not required. On physical examination, they are attached to the skin and cannot be separated from the integument. Treatment is excision of the entire lesion. Drainage of the lesion will temporarily make it smaller, but it will re-enlarge because the cyst wall will continue to produce keratin.

43.4.4 Epidermal Nevus

Epidermal nevi are benign overgrowths of epidermal elements (including keratinocytes); they may be congenital or arise during infancy (► Fig. 43.3). A thick epidermal granular layer, increased basal layer pigment, and inflammatory dermal infiltrate are present. Lesions appear as hyperkeratotic plaques on the extremities, neck, or trunk that may follow the lines of Blaschko. Most are sporadic, but some are associated with epidermal nevus syndromes. Epidermal nevi cause a deformity and pruritus; they do not undergo malignant transformation. Types of epidermal nevi include (1) Becker nevus (late-onset melanosis), (2) comedone nevus (hair follicle nevus), and (3) inflammatory linear verrucous (irritated, pruritic, scaly). Full-thickness removal is performed for symptomatic lesions. Epidermal nevi in unfavorable locations for resection can be improved by shaving of the superficial portion of the lesion, but recurrence is likely.

43.4.5 Granuloma Annulare

Granuloma annulare is a benign, inflammatory condition characterized by ringlike, erythematous lesion (► Fig. 43.4). Histology demonstrates degenerative collagen surrounded by histiocytes, fibroblasts, and lymphocytes. The most common form (localized) is present on the dorsum of the distal extremities. These self-limiting lesions are asymptomatic and typically resolve within 2 years. Large areas can cause disfigurement and reduce the patient's self-esteem. Lesions may be observed or treated with intralesional corticosteroid. Excision can be performed if the diagnosis is unclear or if a problematic lesion does not regress.



Fig. 43.4 A 6-year-old male with granuloma annulare of the foot.



Fig. 43.5 An adolescent male with a keloid of the chest.



Fig. 43.7 A 17-year-old male with a lipoma of flank. Note the lobulated and encapsulated appearance.

43.4.6 Hypertrophic Scar/Keloid

Hypertrophic scars and keloids result from abnormal cicatricial formation. Keloids grow beyond the wound borders and hypertrophic scar does not. Keloids tend to worsen over time, while hypertrophic scars typically improve. Both have a predilection to form on the chest, ear, and shoulder (► Fig. 43.5). Histopathology exhibits fibrous tissue and irregularly arranged dermal collagen. Lesions occur more frequently in dark-skinned individuals. Hypertrophic scars and keloids can be erythematous, painful, and pruritic. First-line nonoperative treatments include pressure, topical silicone, and corticosteroid injection. If these modalities fail, the scar can be excised and the nonoperative therapies can be initiated immediately postoperatively to minimize recurrence.

43.4.7 Juvenile Xanthogranuloma

Juvenile xanthogranuloma is composed of histiocytes and giant cells. It can be congenital (20%) or acquired in early childhood. Lesions initially are pink and transform to a yellow color

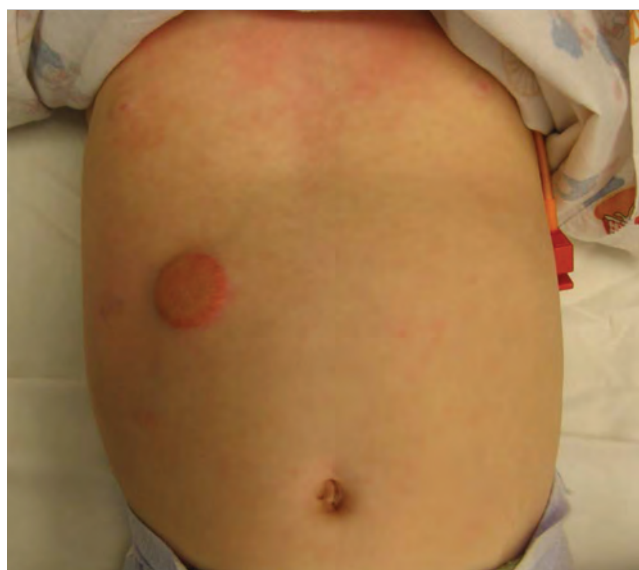


Fig. 43.6 A child with a juvenile xanthogranuloma of the trunk.

(► Fig. 43.6). They may grow rapidly. Juvenile xanthogranuloma can be observed because it has the ability to regress, usually within 3 years. Lesions are difficult to diagnose clinically and thus most undergo excisional biopsy. If the lesion fails to resolve, patients may request removal to improve their appearance.

43.4.8 Lipoma

Lipomas are benign adipose tissue growths of mesenchymal origin. Lesions are subcutaneous with normal overlying skin. They are mobile and may be lobulated (► Fig. 43.7). Lipomas typically are asymptomatic and slow growing. They may be removed for aesthetic reasons or if there is a concern for malignancy. Lipomas are excised through a linear incision over the lesion. Recurrence is minimized by complete extirpation.

43.4.9 Mucocoele

Mucocoeles are oral pseudocysts that result from trauma or obstruction of a minor salivary gland. Lesions typically are fluctuant, mobile, and round. They may have a bluish hue and are most common in the lower lip. Rupture and re-accumulation of mucin can cause size fluctuations. Although mucocoeles may spontaneously resolve, patients often request that they be removed to improve their appearance.

43.4.10 Nevus Sebaceous

Nevus sebaceous is a congenital cutaneous hyperplasia of adnexal elements. Lesions are yellow-orange hairless plaques with a predilection for the scalp and face (► Fig. 43.8). Benign (e.g., trichilemmoma), premalignant (e.g., syringocystoadenoma), and malignant (i.e., basal cell) tumors may develop within nevus sebaceous. The risk of benign or malignant tumor formation is <2%. Nevus sebaceous becomes progressively hyperkeratotic over time (particularly during adolescence) and

thus creates a worsening deformity. Most lesions are removed to eliminate the risk of tumor degeneration and to improve the patient's appearance.

43.4.11 Neurofibroma

Neurofibromas are benign peripheral nerve sheath tumors that contain fibroblasts, mast cells, and Schwann cells. Neurofibromas typically are found in patients with the autosomal-dominant disorder neurofibromatosis type I. A mutation of the NF1 gene on chromosome 17 results in an abnormal tumor suppressor protein called neurofibromin. Patients with neurofibromatosis type I also may exhibit café au lait spots, axillary macules, Lisch nodules of the iris, optic nerve gliomas, and sphenoid dysplasia.

Neurofibromas are categorized as dermal or plexiform. Dermal neurofibromas are soft cutaneous masses or subcutaneous nodules (► Fig. 43.9). They appear commonly during adolescence and remain benign. Most dermal neurofibromas are asymptomatic; resection is reserved for symptomatic lesions causing pain or a deformity.

Plexiform neurofibromas are congenital and cause destruction of soft tissue and bone. They have a 10% risk of malignant

degeneration. MRI can define the extent of the lesion. Removal of these lesions is difficult because they may involve multiple tissue planes and functionally critical nerves.

43.4.12 Pilar Cyst

Pilar (trichilemmal) cysts are keratin-filled lesions surrounded by a wall of stratified squamous epithelium; they may exhibit autosomal-dominant transmission. They originate from the hair follicle outer root sheath and typically occur on the scalp (► Fig. 43.10). In contrast to epidermal cysts, the epithelial lining of a pilar cyst wall lacks a granular layer and lesions do not have a punctum. They typically are concealed by hair but can become infected. Treatment is complete removal of the cyst.

43.4.13 Pilomatrixoma

Pilomatrixoma is a benign calcifying lesion of hair follicle matrix cells. Lesions predominantly affect the head and neck (► Fig. 43.11). It is a slow-growing, firm, fixed mass with a bluish hue. Lesions are usually asymptomatic but can enlarge over time, ulcerate, and become infected. Large pilomatrixomas may cause a visible deformity and psychosocial distress. Pilomatrixomas are excised for definitive diagnosis, to prevent complications associated with their enlargement, and to improve the patient's appearance.

43.4.14 Spitz Nevus

Spitz nevi are benign tumors of spindled and epithelioid melanocytes. These acquired lesions typically exhibit an initial growth period and then remain static. They are reddish brown and usually smaller than 1 cm in diameter. Although Spitz nevi are solitary, they can appear in clusters (agminated) or be diffuse (► Fig. 43.12). Spitz nevi may have histological features similar to melanoma. The Spitz tumor histological spectrum includes Spitz nevus (benign), atypical Spitz tumor (potentially malignant), and Spitzoid melanoma (malignant). Lesions are excised; recommended margins are 2 mm. Treatment of large agminated Spitz nevi can be challenging in areas where complete removal may leave a significant deformity. Localized areas within the agminated lesion can be excised if they change clinically.



Fig. 43.8 A child with nevus sebaceous of the scalp.



Fig. 43.9 A 17-year-old female with neurofibromatosis and a plexiform neurofibroma of the chest.

43.4.15 Verruca Vulgaris

Common warts (verruca vulgaris) are flesh-colored, firm papules that occur from human papilloma virus infection. These growths exhibit hyperkeratosis and acanthosis (► Fig. 43.13). Lesions frequently occur on the hands but can develop on any mucocutaneous surface. Warts are contagious and may be spread by contact. Lesions often resolve spontaneously over a few years, but families typically are eager to treat them because they cause a deformity. Therapeutic modalities include topical medications (e.g., salicylic acid, imiquimod), cryotherapy, curettage, or cauterization. Resection is reserved

for warts that have failed other interventions, but should be performed with caution because the viral infection can recur through the scar.

43.5 Conclusion

Cutaneous lesions are common in children. Although skin growths may appear similar, most can be distinguished by history and physical examination. Excision is reserved for symptomatic lesions, precancerous abnormalities, or to provide definitive histological diagnosis.



Fig. 43.10 A child with pilar cysts of the scalp.



Fig. 43.11 An 8-year-old female with a pilomatrixoma of the brow.



Fig. 43.12 (a) A child with a Spitz nevus of the cheek. (b) A 5-year-old female with an agminated atypical Spitz tumor of the nose.



Fig. 43.13 An adolescent patient with verruca vulgaris on the brow.

43.6 Key Points

- Cutaneous pediatric lesions are common.
- Most disorders can be diagnosed by history and physical examination.
- Management is based on the type of lesion; some mandate resection, while others can be observed.
- Excision is performed to obtain a histopathological diagnosis, treat symptoms, or improve a deformity.

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